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Prevalence of intellectual disability in Finland

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Academic Dissertation

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To my family

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ABSTRACT

BACKGROUND

Intellectual disability (ID) is a developmental condition with problems in mental functions that are reflected in lower than average performance in tests for intelligence, indicating difficulties for the person to adapt in everyday life. The various needs for support of persons with ID necessitate well-organized services systems. To organize, run, and reform those systems reliable, up-to-date, and detailed epidemiological information is needed: What is the prevalence of ID? Where do the persons with ID live? What is their profile of needs of services? Administrative reports from different decades in Finland have indicated that the number of people with ID is around 0.6%. In Finnish population sample studies higher estimates have been reported. International studies have given very variable estimates depending on the population, design, and methods. There is a need to have a more precise estimate of the prevalence of ID in different age groups in Finland.

The aim of this study was to estimate the prevalence of ID in the whole population in Finland, using multiple national health and social-care registers.

METHODS

This study consists of two separate register samples.

In the first sample data were combined in 2000 from eight Finnish national registers, six of which concern benefits connected to long-term illnesses or disabilities allowed by the Social Insurance Institution of Finland (SII) (Child Disability Allowance, Disability Pension, Disability Allowance, Pensioners' Care Allowance, Funding of Rehabilitation, and Preferential Refunding of Long-term Medication), and two concern care provided by hospitals or social welfare (Hospital Discharge Register, and Care Register for Social Care). The list of inclusion diagnoses covered both intellectual disability and those aetiological diagnoses where ID is regularly present (e.g. Down's, Williams', Fragile-X and Angelman's syndromes, and progressive neurological diseases of Finnish heritage). Prevalence estimates were first calculated in four age groups (0–15, 16–39, 40–64, 65+), and thereafter in one-year age cohorts to obtain more understanding of how the prevalence varies with age/year of birth.

The second data set was sampled from Hospital Discharge Registers covering the period 1996–2013 to form an estimate of cumulative prevalence of ID at every age from birth until the maximal age possible (17 years for those born in 1996). The same inclusion list of diagnoses was used.

RESULTS

In the multiple register study, the average prevalence estimate for those of 16–64 years of age was 0.81%. There was a decreasing trend with birth year in this age group, from 0.92% among 64-year-olds to 0.63% among 18-year-olds. Between the ages of 42–52 years the prevalence was exceptionally high, highest at 50 years (1.07%). In old age (65+ years) the average estimate was first 0.38%. At 66 years of age it was 0.49%, decreasing to an average of 0.30% at 80+ years of age. An abrupt drop in the prevalence rate was noticed at 65/66 years of age due to a blind spot in registers and this was corrected by computational means, which yielded a new estimate of 0.75% for those of 65+ years of age. The

validity of the computational correction was evaluated using the register of Preferential Refunding of Long-term Medication.

The Hospital Discharge Register sample yielded a cumulative prevalence of 1.19% at the age of 17. This marked a discontinuity compared with the multiple register study, where the prevalence estimate for the same age was only 0.67%. A total estimate for the population with ID in Finland in 2017 was 53,684 people, which is 0.97%

DISCUSSION.

These multiple register studies suggest a higher prevalence estimate of ID than in most prior register-based studies, but close to estimates found in longitudinal birth-cohort studies. The prevalence given in one-year age cohorts across the whole age span gave a qualitatively different picture and higher prevalence rates than after more robust grouping by age. The cumulative prevalence of ID increases steadily throughout developmental years. Cross-sectional studies at any age do not give a full picture. Inconsistencies in the age-specific prevalence distribution, together with other previous findings, hint at the possibility of so-called hidden disability, i.e. people with difficulties in coping, but not being recognized by the services. The prevalence distribution at all ages reflects in many ways the history of social and health care. There has been a great deal of progress in general, and especially perinatal health. Seasonal epidemics and economic recessions with their implications have affected the opposite direction. The century did also see the great wars of Finland. The emphasis in development of services has fluctuated between segregation and inclusion.

Continuous monitoring of the epidemiology of ID by one-year age cohorts through the most useful registers seems to be both useful and practical. Different registers complement each other. Some reflect diagnostic activities, others, benefits or service delivery. However, for the purposes of service planning, the information in the registers that were used in this study was insufficient. More information would be needed of the actual needs for services, person-by-person. The question arises of whether a specific register of services for intellectual and other developmental disabilities would be helpful or does the information content in current registers develop to provide what is needed.

TIIVISTELMÄ

TAUSTAA

Kehitysvammaisuus on kehityksen myötä ilmenevä mielen toimintojen vaikeus, joka näkyy merkittävästi heikompana suoriutumisenä älykkyyttä mittaavissa standardoiduissa testeissä ja arkisessa toimintakyvyssä. Kehitysvammaiset henkilöt tarvitsevat halki elämän toisten ihmisten apua ja sekä toimivan palvelujärjestelmän. Palvelujärjestelmän rakentaminen, toiminta ja kehittäminen edellyttävät ajantasaista ja yksityiskohtaista epidemiologista tietoa: miten paljon kehitysvammaisia henkilöitä on, missä he ovat ja millaisia ovat heidän palveluntarpeensa. Kansainväliset tutkimukset ovat esittäneet hyvin vaihtelevia lukuja riippuen tutkimuksen asetelmasta ja menetelmistä. Palvelujärjestelmien kehittämisen perustana olleet valtakunnalliset arviot ovat viime vuosikymmeninä esittäneet, että kehitysvammaisia henkilöitä on noin 0,6 % väestöstä. Suomalaiset väestöpohjaiset epidemiologiset tutkimukset ovat päätyneet korkeampiin lukuihin. On tarpeen tarkentaa arviota kehitysvammaisuuden esiintyvyydestä Suomessa.

Tämän tutkimuksen tavoitteena oli arvioida kehitysvammaisuuden esiintyvyyttä Suomen koko väestössä nojautuen valtakunnallisiin sosiaali- ja terveydenhuollon rekistereihin.

AINEISTO JA MENETELMÄT

Tämän tutkimuksen aineisto koostu kahdesta eri poiminnasta.

Ensimmäinen poiminta suoritettiin monirekisteritutkimuksena vuonna 2000 kuutta KELAn etuusrekisteriä (lapsen hoitotuki, työkyvyttömyyseläke, vammaistuki, eläkkeensaajan hoitotuki, kuntoutus ja erityiskorvattavat lääkkeet) sekä kahta THL:n rekisteristä (sairaaloiden hoitoilmoitusrekisteri ja sosiaalihuollon hoitoilmoitusrekisteri) hyödyntäen. Poimintadiagnoosien luettelo koostui kehitysvamman tasodiagnoosien lisäksi myös syydiagnooseista, joiden oirekuvaan säännönmukaisesti liittyy kehitysvammaisuus (esimerkiksi Downin, Williamsin, Angelmanin ja Fragile-X oireyhtymät ja suomalaisen tautiperintöön lukeutuvat etenevät sairaudet). Kehitysvammaisuuden esiintyvyys laskettiin aluksi neljälle ikäryhmälle (0–15, 16–39, 40–64 ja 65–) ja sen jälkeen ikävuosikohorteittain, jotta voitiin tarkemmin tutkia esiintyvyyden vaihtelua iän/syntymävuoden mukaan.

Toinen poiminta kerättiin terveydenhuollon hoitoilmoitusrekisteristä 1996–2013 käyttäen samaa poimintadiagnoosien luetteloa. Tästä aineistosta laskettiin kumulatiivinen esiintyvyys syntymästä alkaen siihen asti, kuin aineisto salli (vuonna 1996 syntyneiden osalta 17-vuotiaaksi).

TULOKSET

Monirekisteritutkimuksen perusteella kehitysvammaisuuden keskimääräinen esiintyvyys 16–64-vuotiaiden ryhmässä oli 0,81 %. Esiintyvyys tässä ikäryhmässä laski tasaisesti syntymävuoden myötä. Vuonna 1936 syntyneillä (64-vuotiailla) se oli 0,94 % ja vuonna 1982 syntyneillä (18-vuotiailla) vastaavasti 0,63 %. Ikäryhmässä 42–52 vuotta esiintyvyyksluvut olivat poikkeuksellisen korkeita, korkein esiintyvyys oli 50-vuotiaiden ikäryhmässä (1,07 %). Yli 65-vuotiaiden ikäryhmässä keskimääräinen esiintyvyys oli ensin 0,38 %. 66-vuotiaiden keskuudessa esiintyvyys oli 0,49 % laskien 80 vuotta täyttäneiden kohdalla 0,30 %:iin. Mahdollisesta rekisterikatveesta johtuen suoritettiin korjauslaskelma, jonka luotettavuutta arvioitiin KELAn erityiskorvattavien lääkeoikeuksien rekisterin avulla. Korjauslaskelman tuottama uusi esiintyvyyssarvio oli 0,75 %. Sen luotettavuutta arvioitiin

erityiskorvattavien lääkeoikeuksien rekisterin avulla. Sen tuottama uusi esiintyvyyssarvio yli 65-vuotiaille oli 0,75 %.

Hoitoilmoitusrekisteripöiminnan perusteella kehitysvammaisuuden esiintyvyys 17-vuotiaiden ikäryhmässä vuonna 2013 oli 1,19 %. Tulos poikkesi huomattavasti monirekisteritutkimuksen tuloksesta, joka samalle ikäryhmälle oli 0,67 %. Tutkimusten nojalla voidaan arvioida, että vuonna 2017 maassamme asui 53 684 kehitysvammaista henkilöä (0,97 % väestöstä).

POHDINTA

Monirekisteritutkimuksen tulokset ovat korkeampia kuin useimmissa aiemmissa rekisteripohjaisissa tutkimuksissa ja lähestyvät syntymäkohorttien pitkittäistutkimuksia. Ikävuosittain koko ikäskaaan osalta esitetyt esiintyvyyshlutut tarjoavat laadullisesti erilaisen kuvan ja korkeampia esiintyvyyshlukuja kuin karkeammat ikäluokittain esitetyt tulokset. Kumulatiivinen esiintyvyys kasvaa tasaisesti koko kehitysiän. Poikittaistutkimukset missä tahansa iässä eivät anna kokonaiskuvaa. Epäjohtonmukaisuudet ikävuosikohtaisessa jakaumassa yhdistettynä aiempien tutkimusten tuloksiin viittaavat mahdollisuuteen, että väestöstä jää tunnistamatta kehitysvammaisia, jotka olisivat palveluiden tarpeessa.

Ikävuosikohtaiseen esiintyvyyshjakaumaan heijastuu monin tavoin jopa sadan viime vuoden sosiaali- ja terveydenhuollon historia. Siihen kuuluvat terveydenhuollon merkittävät edistysaskelet, ja päinvastaiseen suuntaan kausittaiset epidemiat ja taloudelliset lamat seurausvaikutuksineen. Jaksoon mahtuvat myös Suomen kokemat suuret sodat. Etuus- ja palvelujärjestelmän kehittämiseen on puolestaan sisältyntä kausia, jolloin on painotettu erityispalveluita tai päinvastoin niitä kartettu.

Kehitysvammaisuuden esiintyvyyden jatkuva seuranta ja kuvaaminen yksivuotiskohorteittain keskeisten rekisterien tarjoaman informaation kautta näyttäisi olevan sekä hyödyllistä että käytännöllistä. Eri rekisterit täydentävät toisiaan – toiset heijastavat diagnostisia tutkimuksia, toiset taas palveluiden/etuushien saajia ja tarjontaa. Palveluiden kehittämisen tarpeisiin tässä käytettyjen rekistereiden tietosisältö ei kuitenkaan ole riittävää. Enemmän tietoa tarvitaan yksittäisten kehitysvammaisten henkilöiden todellisista tarpeista. Herää kysymys, tarvittaisiinko erityinen kattava palvelurekisteri henkilöille, joilla on kehitysvamma tai muu kehityksellinen erityispalveluiden tarve vai kehittyvätö olemassa olevat rekisterit niin, että niistä tärkeä tieto on riittävän hyvin saatavissa.

ABBREVIATIONS

AAIDD	the American Association on Intellectual and Developmental Disabilities
AAMR	the American Association on Mental Retardation
AB	adaptive behaviour
AF	adaptive functions/functioning
avg	average
BIF	borderline intellectual functioning
CARE	Care Register for Social Care
CDA	Child Disability Allowance
CI	confidence interval
DA	Disability Allowance
DP	Disability Pension
DSM	Diagnostic and Statistical Manual (of Mental Disorders)
f	female(s)
GP	general practitioner
HOSP	Hospital Discharge Register
ICD	International (Statistical) Classification of Diseases (and Related Health Problems)
ICF	International Classification of Functioning, Disability, and Health
ID	intellectual disability
IQ	intelligence quotient
LAMI	low and middle income (countries)
m	male(s)
MBD	minimal brain dysfunction
MED	Preferential Refunding of Long-term Medication
mID	mild intellectual disability
NIDD	the National Intellectual Disability Database (in Ireland)
PCA	Pensioners' Care Allowance
PCMR	President's Committee on Mental Retardation
PIC	personal identity code
REH	Funding of Rehabilitation
SD	standard deviation
SEM	standard error of measurement
SES	socioeconomic status
sID	severe intellectual disability
SII	The Social Insurance Institution of Finland
SMR	standardized mortality rate
SSC	social security code
STAKES	National Research and Development Centre for Welfare and Health (later THL)
THL	National Institute for Health and Welfare
WAIS	Wechsler Adult Intelligence Scale
WHO	World Health Organization
WISC	Wechsler Intelligence Scale for Children
y	year(s)

EPIDEMIOLOGICAL TERMS RELATED TO STUDIES OF INTELLECTUAL DISABILITY (ID)

Incidence (or incidence rate)	Incidence (or incidence rate) is the number of new cases per population at risk in a defined period. When the period is long, the incidence becomes cumulative. The incidence of ID is not the same throughout developmental years. Some syndromes can be diagnosed prenatally and others immediately or soon after birth, or later up to 18 years (Wilska and Kaski 1999). Often incidence of ID is a confusing term, e.g. when there is a slowly emerging lag in development due to some biological predisposition. In such cases it is more a question of when to decide that the cognitive development and concomitantly adaptive skills are reliably lagging the norms. Aetiological investigations, which are then considered fit for purpose, may reveal an inborn syndrome. In such cases, the condition has emerged at conception (or even before), and the incidence figure is an administrative one.
Cumulative incidence	Cumulative incidence is calculated by the number of new cases during a period divided by the number of subjects at risk in the population at the beginning of the study. In other words, deceased persons are not excluded, neither from the cases nor the risk population. For conditions which practically never resolve, like ID, cumulative incidence tells what the prevalence would be if no deaths had occurred. The difference between cumulative incidence and prevalence at a certain age depends on mortality rate.
Prevalence	Prevalence is the proportion of cases with a disease/condition in a population.
Point prevalence	Point prevalence is the proportion of a population that has the condition at a specific point in time.
Period prevalence	Period prevalence is the proportion of a population that has the condition at some time during a given period (e.g., 12-month prevalence), and includes people who already have the condition at the start of the study period as well as those who acquire it during that period. The period can be the whole lifetime.
Cumulative prevalence	Cumulative prevalence is used here for clarity on occasions where the prevalence at certain time points has been reconstructed from cases tracked cumulatively from a time before and all cases are still alive. ID which has reliably been diagnosed is a condition that practically never resolves. Thus, this kind of reconstruction is possible.
Psychometric prevalence	In studies of ID the prevalence rate which relies solely upon intelligence tests, without considering adaptive skills, is called psychometric prevalence.
Administrative vs. true prevalence	Administrative prevalence is based on the known cases, mostly recognized by services. True prevalence is the theoretical figure considering also those who are not identified.
Mortality rate	Mortality rate is the proportion of individuals dying in the risk population during a defined period.
Life expectancy	Life expectancy is the average time a person is expected to live, based on the year of birth and other demographic factors including gender, assuming that the current mortality rates remain unchanged.
Standardized mortality ratio	The standardized mortality ratio is the ratio of observed deaths in the study group to expected deaths in the general population.
Migration	Migration means individuals moving into the study population (immigration) or out of it (emigration). In a cohort study, where the study population is defined by individuals at the beginning, immigration cannot happen, but emigration can mean loss of cases. In contrast, in a study where the risk population is not defined on an individual level, but is, for example, persons living in an area, migration may change the prevalence of ID.

LIST OF ORIGINAL PUBLICATIONS

- I. Westerinen H, Kaski M, Virta L, Almqvist F, Iivanainen M (2007). Prevalence of intellectual disability: a comprehensive study based on national registers. *Journal of Intellectual Disability Research* 51(9):715-725.
- II. Westerinen H, Virta L, Kaski M, Almqvist F, Iivanainen M (2014). Age-specific prevalence of intellectual disability in Finland at the beginning of a new millennium – multiple register method. *Journal of Intellectual Disability Research* 58(3):285-295.
- III. Westerinen H, Kaski M, Virta LJ, Almqvist F, Kautiainen H, Iivanainen M (2016). The prevalence of intellectual disability among one-year cohorts of Finnish elderly as determined with the multiple register method. *Journal of Intellectual & Developmental Disability* 41(3):267-271.
- IV. Westerinen H, Kaski M, Virta LJ, Kautiainen H, Pitkala KH, Iivanainen M (2017). The nationwide register-based prevalence of intellectual disability during childhood and adolescence. *Journal of Intellectual Disability Research* 61(8):802-809.

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1 INTRODUCTION

Intellectual disability (ID) is a developmental condition with problems in mental functions that are reflected in lower than average performance in tests for intelligence, indicating difficulties for the person to adapt in everyday life (Switzky and Greenspan 2006). Intellectual disability is a human condition that directly or indirectly concerns most people. Although rare, about one in a hundred have ID (Maulik et al. 2011). This means that many of us have persons with ID as relatives, many as neighbours, or we just occasionally come into contact with one, just enough to feel compelled to think and feel what it means. It is one of the many conditions, like physical or sensory handicaps and mental disorders, which can bring many kinds of challenges in life. Somatic comorbidities with ID are multiple, very frequent, and often painful/disabling (Kinnear et al. 2018). In the most severe forms the very meaning of existence may be questioned if no contact develops between the person and others. In milder forms social interactions develop, which may bring joy to both parties. In lucky situations, the person will live a normal life.

The prevalence of ID varies according to populations, research methods, and the definition of ID (McLaren and Bryson 1987, Roeleveld et al. 1997, Maulik et al. 2011). The prevalence of severe ID is relatively constant across studies (Abramowicz and Richardson 1975, Roeleveld et al. 1997), but milder forms are not so well identified. However, it would be important to know the real prevalence. The various needs for support of persons with ID necessitate well-organized service systems. To organize, run, and reform those systems, reliable, up-to-date, and detailed epidemiological information is needed: What is the prevalence of ID? Where do the persons with ID live? What is their profile of needs of services? (Anagnostopoulos and Soumaki 2011).

A proportion of cases of ID is caused by known preventable medical conditions (infectious diseases, environmental toxins, congenital hypothyreosis, foetal alcohol syndrome etc.). It is important to follow-up their incidence and study the impact of prevention programmes (Fryers and Mackay 1979, Bower et al. 2000, McKenzie et al. 2016).

Another part of ID is caused by medical disorders that are not yet well known. Studying their epidemiology may help to reveal their aetiology and pathogenesis, and open new possibilities for prevention and/or management.

Epidemiological research is needed for comparison of different populations to find risk factors and possible aetiologies, and to compare service systems between countries (Holt et al. 2000).

In the present studies we explored the prevalence of ID in the whole population of Finland at various ages, using different sets of register-based data.

2 REVIEW OF THE LITERATURE

2.1 HISTORICAL DEVELOPMENT OF THE DEFINITION OF ID

Understanding of the concept of ID has changed along with the development of science, as medical aetiologies have replaced magical thinking (Scheerenberger 1983). On the other hand, the sociocultural dimension is ever-present, which means how differences between people are valued, appreciated or tolerated. Psychological science is learning and teaching how, despite problems in neurological development, the development of the individual mind can be supported. Thus, ID is both a collection of medical backgrounds and a profoundly human and cultural condition (Sinason 1985, Rapley 2004).

Several themes have evolved over the last one hundred years. Research in medicine has characterized specific syndromes that regularly or always lead to ID (see *Table 1*), and there is a multitude of others which pose a variable risk to intellectual development; nowadays subtler genetic variations than gross syndromes (Amberger et al. 2011). The term behavioural phenotype denotes the fact that a psychological developmental line may be typical of the genetic background (Di Nuovo and Buono 2011, Reilly 2012).

In addition to the above, psychological understanding has developed from undifferentiated descriptions of mind/mental functions to the first emphasis of different facets of intelligence and its measurement, then to more detailed neuropsychological assessment (Burack et al. 1998). The crucial role of adaptive abilities/functions/behaviour apart from intelligence has become a major part in the definition and assessment. Social underpinnings, social construction of identity and abilities have challenged the too-narrow medical description of ID (Rapley 2004).

Table 1. Some medical syndromes which regularly or always lead to ID, the year they were defined, and researchers who discovered the condition (from various sources)

1866 Down's syndrome by Sir Langdon Down
1880 Tuberous sclerosis complex by Désiré-Magloire Bourneville
1934 Phenylketonuria by Ivar Asbjørn Følling
1956 Prader-Willi syndrome by Andrea Prader, Alexis Labhart, and Heinrich Willi
1959 Trisomy 21 identified as the underlying genetic abnormality in Down's syndrome by Marthe Gautier
1961 Williams' syndrome by John C. P. Williams
1967 Aspartylglucosaminuria by F.A. Jenner and R. J. Pollitt
1968 Foetal alcohol syndrome originally described by P. Lemoine and colleagues
1973 Cohen's syndrome by Michael Cohen
1981 Velocardiofacial syndrome by Robert Shprintzen and colleagues
1983 Rett's disorder by Andreas Rett
1985 Fragile-X syndrome by Felix F. de la Cruz

Several organizations have published diagnostic manuals on ID. The American Association on Intellectual and Developmental Disability (AAIDD) (originally the American Association for the Study of the Feeble-minded, then the American Association on Mental Retardation, AAMR) has published

eleven revisions of its manual over the period of 1919–2010. The American Psychological Association (APA) has included ID into its Diagnostic and Statistical Manual of mental disorders (DSM), the fifth of which (DSM5) was published in 2013. The WHO has published succeeding versions of the International Classification of Diseases (ICD), which also includes ID in the section on mental disorders – the tenth version is now in use, and the eleventh in preparation.

2.1.1 AAMR/AAIDD

The development of definitions of ID can be followed in succeeding revisions of the AAMR/AAIDD manuals.

In the first versions of the manuals, developmental lag was expressed somewhat superficially (Luckasson et al. 2002). Development of suspected persons was compared with that in normally developing children. In 1905 psychometric tests were developed and in 1912 they were introduced in the USA specifically to diagnose persons with ID. Once intelligence quotient (IQ) tests were introduced, definitions tended to be in terms of measured intellectual ability (Whitaker 2013). The 1959 edition defined a cut-off point at one standard deviation (SD) below the population mean of the respective age group (Heber 1959). In the 1973 revision the cut-off point was lowered to -2 SD (Grossman 1973), but due to measurement error in 1983 it was set more loosely as $-1\frac{2}{3}$ – -2 SD (Grossman 1983), which has prevailed in the following revisions. A disproportionate number of people from lower social classes and ethnic groups were considered to be intellectually disabled. It was realized that the reason for this over-inclusiveness of the diagnosis was that it was entirely based on IQ (Whitaker 2013). This was the main reason for laying more emphasis on the second criterion – adaptive skills.

Similarly, adaptive behaviour was first expressed in general terms, and later more exact operational terms were introduced. In 1908 a person with ID was described as "unable to perform his duties as a member of society in the position of life to which he is born" (Luckasson et al. 2002). In 1959, the definition of ID included for the first time the adaptive behaviour criterion, impairment in one or more of the following: maturation, learning and social adjustment (Heber 1959). In 1973, the adaptive behaviour criteria were set according to age groups: in early years, sensorimotor skills, communication, self-help, socialization; in childhood and early adolescence, application of basic academic skills in daily life, application of reasoning and judgement; in later adolescence and adult life, vocational and social responsibilities and performances (Grossman 1973). In 1992 adaptive skills were separated into ten areas: communication, self-care, home living, social skills, community use, self-direction, health and safety, functional academic skills, leisure and work (Luckasson et al. 1992). Limitations needed to be present in two or more areas. The 2002 revision stated that limitations in adaptive behaviour should be established using standardized measures. The nine areas were replaced with three types of adaptive behaviour: conceptual, social, and practical. To fulfil the criteria of ID, performance should be under two standard deviations below the mean, either in one or more of the areas or in overall score (Luckasson et al. 2002).

Expression of the age criterion in the diagnosis of ID has changed in succeeding versions of classification systems. Constantly the main emphasis has been on the developmental aspect – the condition of ID either originates (Heber 1959) or becomes manifest (Grossman et al. 1973) during the developmental period. Later, the developmental period was fixed to some defined age, up to approximately 16 years (Heber 1961), later to 18 years (Luckasson et al. 1992). Although the age

criterion refers to the developmental period, it is not based on the modern view of development. Maturation of the brain and concomitant psychological development continues well into the third decade of life (Brenhouse and Andersen 2011). With cognitive impairment the developmental tasks of late adolescence and early adulthood, such as gaining independence from parents may be postponed until later, which makes a strict age criterion (16 or 18 years) problematic (Zetlin and Morrison 1998).

An important broadening of scope was to focus on an individual's social context, as opposed to person-based assessment. The theory of social construction has shown how persons categorized as intellectually disabled are formed, as such, in and through their moment-by-moment interaction with care staff and other professionals and people around (Rapley 2004). The focus on social underpinnings, the environmental context, has been given greater weight since the 1992 revision, in addition to the traditional criteria of IQ and adaptive skills (Luckasson et al. 1992). Environmental conditions and supportive structures have been considered independent or intervening variables, whereas a person's functioning level, living/employment status, and level of satisfaction are dependent variables. In the 2002 revision the emphasis on context was made still stronger, and its components and interactions analysed (Luckasson et al. 2002). It has also been noted that the assessment of context is not typically accomplished by means of standardized measures (like intelligence, and adaptive behaviour), but it is a necessary component of clinical judgment and integral to understanding the individual's functioning.

This short review of succeeding editions/revisions of manuals by AAMR/AAIDD does not do justice to the broad discussion and wide-ranging ideological development behind the sometimes minor changes in wording.

2.1.2 DSM

Successive versions of the Diagnostic and Statistical Manual of psychiatric disorders by the American Psychiatric Association have also shown major transformations in the definitions of ID. DSM-II did not emphasize IQ cut-off scores (American Psychiatric Association 1968). Previous versions contained outdated and now offensive terminology (such as 'idiot' and 'imbecile') (Greenspan and Woods 2014). DSM-IV-TR was largely based on the definition of adaptive functioning proposed by the AAIDD (Luckasson et al., 1992). It defined adaptive functioning deficits as concurrent impairments (e.g., performance approximately 2 SD below the mean) in at least two theoretically derived adaptive skill areas (i.e., communication, self-care, home living, social/interpersonal skills, use of community resources, self-direction, functional academic skills, work, leisure, health, and safety) (American Psychiatric Association, 2000).

In DSM-5 (American Psychiatric Association 2013) adoption of the term intellectual developmental disorder (IDD) represented a shift from a disability (test score) emphasis to a disorder (medical/neurobiological) (Greenspan and Woods 2014). This change in DSM-5 was a culmination of various efforts to broaden the diagnosis to include individuals at the upper range of impaired scores, who had been prevented from receiving the diagnosis because of overly rigid reliance on arbitrary IQ cut-off scores (Greenspan and Switzky 2003). Operationally the shift was expressed by elevating the upper IQ ceiling for ID to 75, considering the Flynn effect (see Chapter 2.1.5.3), noting that highly discrepant individual subtest scores may make an overall IQ score invalid, and considering replacing IQ tests with more informative neuropsychological testing to approach executive functions and other dimensions of cognition. In DSM-5 the new criteria require impairment in one adaptive domain rather than two or more skill areas. Papazoglou et al. (2014) noted that the new criteria led to diagnosis of 9%

fewer individuals. Tassé et al. (2016) criticized the wording in DSM-5, implying a causal link between intellectual functioning and adaptive behaviour.

2.1.3 WHO – International Classification of Diseases (ICD)

The WHO has given a definition that, interestingly, has a slightly different emphasis (World Health Organization 1996). It defines ID as a condition of arrested or incomplete development of the mind, which is especially characterized by impairment of skills manifested during the developmental period. These skills are considered to contribute to the overall level of intelligence, i.e. cognitive, language, motor, and social abilities. Although operationalization of the ICD definition approaches that of AAMR/AAIDD, it is the only definition that mentions 'the mind'. Later, the WHO also adopted a definition which is in line with the AAIDD definition, and stress is on cognitive aspects: "Intellectual disability means a significantly reduced ability to understand new or complex information and to learn and apply new skills (impaired intelligence). This results in a reduced ability to cope independently (impaired social functioning), and begins before adulthood, with a lasting effect on development." (World Health Organization 2018.)

The WHO also produced another manual to be used alongside the ICD classification, originally the International Classification of Functioning, Disability, and Handicaps (ICIDH) (World Health Organization 1980). The primary goal was to provide a tool for epidemiological research and outcome-related evaluations of care systems on a national and international level. Its basic assumption of the sequence "etiology → pathology → manifestation" was conceptually criticized, mainly because of the unidirectional and allegedly causal nature of the disabling process, and absence of the impact of the environment. The revision was developed in 1993–2001, with the official release of the International Classification of Functioning, Disability, and Health (ICF) in 2001 (World Health Organization 2001). Its conceptual model has become close to that of the IAADD 2002 revision (Buntinx 2002).

In Finland, the ICD classification system is used in health care. However, the AAMR/AAIDD manuals have influenced both research and clinical practice. For example, the first large Finnish epidemiological population sample study of ID conducted in 1962 (Ruoppila 1966) made reference to the then recently published edition of AAMR manual (Heber 1959).

2.1.4 Considerations on terminology

The introduction of the term *intellectual disability* has largely replaced various earlier terms such as *mental retardation*, *mental deficiency*, *feeble-mindedness*, *mental subnormality* or even older Latin- or Greek-based terms such as *oligophrenia*, *idiocy*, *imbecility*, and *moronity*. The new and nowadays recommended term ID emphasizes intellect but leaves aside the word *mental*. Mental refers to mind, which is a broader concept than intellect and cognitive functions (Scheerenberger 1983).

The word 'retardation' emphasizes the delay in development, which is true: almost always a person with ID will develop but reach (part of) the developmental milestones at a later age. The phrase ID shifts the focus to the consequences of this delay, i.e. various difficulties in functioning. *Mental handicap*, also a formerly used term, implied both these connotations. *Cognitive impairment* is sometimes used to refer to the supposed central role of intelligence in developmental disability. Greenspan, in contrast, has stressed the role of effects in development, maturation, and intellectual development (Greenspan 1979, Greenspan and Benderly 1997, Greenspan 2001). In maturation of the mind, intellect has an important

role – on the other hand intellect is an important result of maturation, which is highly dependent on early social interaction (Greenspan and Benderly 1997).

In earlier times behavioural deviance and physical deformity were important defining characteristics of what we now call ID (Scheerenberger 1983). The central role of intellect in the definition and classification of ID was historically related to the interest in measuring cognitive functions, starting in the first decade of the 20th century. This development was intended to bring scientific rigor, with numeric values. However, the rather broad idea of intellect was for a long time narrowed down to certain aspects that the developed tests measure, such as verbal and visual reasoning. These measures predict how well students do at school, what kind of job they will later obtain, and how well they perform that job. Nevertheless, the existence of multiple types of intelligence has been proposed, and *practical intelligence*, for example, has been suggested to exist independently of *academic intelligence* (Mackintosh 2011). This development of thinking was mirrored in the demand to include *adaptive abilities* in the definition of ID.

In multicultural contexts these phenomena seem to become even more complex (Jenkins 1998). Although in all societies some distinction is made between competence and incompetence, there is enormous variability concerning where the line is drawn and what it means. A wider notion of mind and mental capacity is considered more relevant for understanding the situations of 'incompetent' people in everyday life. Mental capacity is a broader term than intelligence, referring to will, intention and feelings etc. (Whyte 1998).

Nowadays intellectual disability is often studied jointly with other developmental disabilities (especially autistic spectrum disorders), using the term IDD, intellectual and developmental disabilities (Emerson 2012). Although the conditions have different features and diagnostic criteria, they broadly overlap in problems with coping and service needs (Luckasson et al. 2002).

In the Finnish language, the dominant term for ID is "kehitysvammaisuus", the direct translation being developmental disability as abbreviated from "älyllinen kehitysvammaisuus", intellectual developmental disability. In Finland the term "kehitysvammaisuus" (developmental disability) has been established to refer solely to ID.

The role of cut-off values of IQ or measures of adaptive behaviour in diagnosis has several connotations. Scientific studies, both epidemiological and intervention studies, require a reliable definition of the study population. For an individual it means labelling, which may have positive or negative connotations, largely depending on the family, and cultural values. Administratively a cut-off value in diagnosis determines those who are eligible to receive support intended for persons with ID. And finally, concerning a small minority, but with dramatic consequences, in certain states in the USA it may determine whether a capital punishment is executed or not (Trahan et al. 2014).

2.1.5 Measurement of intellectual performance

2.1.5.1 Various tests

Most definitions of ID refer to significantly sub-average intellectual functioning as one of the diagnostic criteria. Intelligence, in terms of functioning, is "a general mental ability, which includes reasoning, planning, solving problems, thinking abstractly, comprehending complex ideas, learning quickly, and

learning from experience." Although there is strong support for the concept of "general intelligence" or "g", there is no gold standard to be used as a reference, and different developed tests have only partial correlations with each other (Luckasson et al. 2002).

Typically, the tests measure performance in several major dimensions. The Wechsler Intelligence Scale for Children (WISC) has six subtests for verbal intelligence and six for perceptual-motor abilities (Wechsler 1991). The Stanford-Binet test battery has scales for reasoning ability, quantitative reasoning ability, abstract/visual reasoning ability, and short-term memory (Thorndike et al. 1986). The Cognitive Assessment System, based on a multidimensional assumption of intelligence, contains four scales, Planning, Attention, Simultaneous, and Successive (Naglieri and Das 1997). In addition to the tests which are used also for the general population, there are tests for special circumstances, such as for infants (Bayley Scales) or non-speaking persons (Leiter-R), or a test to be used in a short time (the Slosson Intelligence Test) (Luckasson et al. 2002).

Originally the tests (such as Wechsler's) were not intended to be used in the low-ability range, more than two SDs below average (Luckasson 2002). Standardization samples rarely include an adequate number of subjects with ID needed to provide sensitive measurement in the very low ability range (Hessl et al. 2009).

Different tests identify different proportions of individuals as potentially having ID, with the same cut-off of -2 SD – the difference may be as high as 2.28% vs. 3% (Luckasson 2002). It has also been noted that the WISC-III classified disproportionately more Blacks than Whites as having ID as compared with the Cognitive Assessment System (Naglieri and Das 2001).

In epidemiological studies of ID, many different tests for intelligence have been used. In a single study, results from up to nine different tests might have been used variably for different subjects (Heikura et al. 2003).

2.1.5.2 Cut-off values, standard error of the tests, and levels of ID

The IQ cut-off value has a great influence on the prevalence of ID over time. According to theoretical normal distribution the prevalence would be 1.94% at $IQ < 70$, or 2.28% if the cut-off is set to ≤ 70 as in several studies (i.e. < 71 , with a difference of just one point). Widening the range to 69 ± 2 points yields a prevalence range of 1.39% – 2.66%. If the upper limit is extended to 75, as recommended in the 8th edition of the manual by AAIDD onwards as a result of the standard error of tests (Grossman 1983), the theoretical prevalence of ID would be as high as 4.78%.

Consideration of the standard error of measurement (SEM) is important. For example, in WISC-III, one of the common tests for intelligence, the SEM for full-scale IQ is 3.20. For interpretative purposes, one can be 95% confident that an individual's true score falls within ± 1.96 SEMs. Thus, an individual whose tested IQ is 65 has a true IQ roughly somewhere between 59 and 71 (Luckasson et al. 2002).

Levels of ID have been defined mainly in two ways. The more precise way in the main classification systems (AAMR/AAIDD, DSM, ICD) divides ID into four classes: mild, moderate, severe and profound. The other way, used in many epidemiological studies, is to keep mild (mID) and combine the other three as 'severe'. In this text severe ID (sID) refers to the latter, unless otherwise specified. The criteria for these classes have also varied. Besides the changing criteria for the upper IQ limit of mID, the IQ cut-off values between the classes have varied. For this literature review, the IQ cut-off value

between mID and sID is most important. It has been set to 50–55 points. Theoretically, in a normal distribution, more than 95% of persons with an IQ of <70 should be included in the mID group. However, it has been reasoned that in addition to the lower end of the normal distribution of intelligence, specific syndromes add prevalence, especially at the lowest end (Whitaker 2013). That may be the reason why the prevalence of sID is many times greater than expected from a normal distribution (the probability of $\text{IQ} \leq 50$, with average 100 and SD 15 in a normal distribution is 0.04%). Even so, in most of the published studies the proportion of cases of mID has been much lower than expected. In such cases, one can reason that either the screening has not been comprehensive, or in the assessment adaptive behaviour has compensated for low IQ values in the final diagnosis.

2.1.5.3 *Flynn effect*

The Flynn effect denotes the continuous increase in intelligence test scores over time at the population level (Flynn 2007). It was first noted in Scotland when comparing group IQ test results on virtually the entire population of 11-year-olds in 1933 and 1947 (Mackintosh 2011). The increase was some 2–3 IQ points between the time points. The results were confirmed in Leicester between 1936 and 1949, where an increase of 1.3 IQ points was noted in schoolchildren (Mackintosh 2011). Subsequent research was documented and summarized by Flynn. He noticed that the true rate of increase in test scores was higher than thought (Flynn 2007). The current estimate of inflation of test norms is 0.3 points per year. The most recent research has suggested that the increase is slowing down or has ended, at least in some countries with relatively high average national IQs. The increase has not been the same in different subtests. For example, among the various tests from the Wechsler Intelligence Scale for Children (WISC) from 1949 to 2002, the gains in test points were large (15 to 27 points) in connection with Raven's Matrices, Similarities, and Block design, but very small (3 points) in vocabulary, information and arithmetic tests (Flynn 2007).

The implications of the Flynn effect are actually very challenging: assessed by the current IQ test norms, the average person would have been scored at the borderline or mild ID level one hundred years ago. There are many suggestive explanations for the Flynn effect, such as improved childhood nutrition and health, universal education, smaller families and the influence of educated mothers on their children, test sophistication, and demographic changes (Mackintosh 2011). It has also been suggested that abstract thinking, and categorizing has become more valued than more organic association between things. Flynn (2007) has given an example of how our thinking has changed. When presented with a Similarities-type item such as "what do dogs and rabbits have in common?", Americans in 1900 would have been likely to say, "You use dogs to hunt rabbits." A high-scored answer would be that they are both mammals, although that is less interesting, as Flynn points out. One can ask how much this can be considered to represent higher intelligence or just another way of thinking about the world around.

Recently, reversal or slowing down of the Flynn effect has been observed in some countries (Dutton et al. 2016). In an analysis of Norwegian birth cohorts from 1962 to 1991, both the Flynn effect and its reversal seemed to be more environmentally than genetically caused (Bratsberg and Rogeberg 2018).

2.1.5.4 *Setting norms in the study population*

The definition of ID is confusingly relativistic. While it is seemingly exact and scientific – performance (both cognitive and adaptive) about 2 SD less than average – serious problems appear concerning the norm setting. If the norms were set in every study population, there would be very few differences in the prevalence of ID. An epidemiological study would only serve the purpose of finding the persons

with ID in a study population, and what characteristics they have. However, local norms have been used to give emphasis to specific features of the study population, such as ruling out global intellectual delay when identifying primary language disorder in children from minority populations (Lancaster and Camarata 2016) or limiting the misidentification of reading disabilities in lower socioeconomic community populations (Malliet 2015). In contrast, if the tests are not normalized in the study population, which norms should be used? How much are the norms culturally dependent? Is it so that in any population, those who score 2 SD units below the average, for example, need the same level of support to cope? Or is there some universal absolute level of intellectual performance to which ability to cope is associated?

The sizes of populations used in standardization have been around some thousand individuals – the Wechsler Intelligence Scale for Children-III (WISC-III) was normed on a sample of 2200 children, and the Wechsler Adult Intelligence Scale-III (WAIS-III) was standardized on 2450 adults in the United States. If the study population is much smaller, the standard error of measurement will increase from that of the original standardization sample, e.g. 3.20 points for WISC-III (Luckasson 2002).

There are practical problems impeding the normalization of tests in a study population. First, normalization can only be performed in a large representative population sample, which means the study must be population sample. Second, testing large groups requires costly resources. In practice the norms are set in national samples, often repeatedly, time after time.

2.1.6 Measurement of adaptive behaviour

2.1.6.1 Various methods

The introduction of measures of intelligence dates to the early 1900's, but the concept of social competence was not formally recognized until Edgar Doll proposed measuring an individual's social maturity some thirty years later (Paskiewicz 2008). Adaptive behaviour (AB) is a multidomain construct. Scales that have been developed place different degrees of emphasis on various domains. The most widely used and best-validated instruments contain the domains mentioned in the current revision of the AAIDD manual – conceptual skills, social skills, and practical skills

IQ is measured by means of standardized tests, but defining the level of adaptive skills needs observation in natural settings or interviews with those who can give a reliable report. Keith et al. (1987) clarifies the relationship between constructs: (1) intelligence is conceptualized as a thought process, whereas adaptive behaviour emphasizes everyday behaviour; (2) intelligence scales measure maximum performance, whereas adaptive behaviour scales measure typical performance; and (3) intelligence scales assume a stability in scores, whereas when using adaptive behaviour scales it is assumed that performance can be modified. This has been noticed, for example, when a person is given new possibilities in more stimulating environments after closure of an institution (Saloviita 2009). Adaptive behaviour is considered to exist and develop (partly) independently of intellectual functions (Tassé et al. 2012). However, there is still much uncertainty concerning whether or not adaptive behaviour is a coherent construct (Whitaker 2013).

The idea of a cut-off at 2 SD units below the mean in adaptive behaviour as another criterion of ID was introduced into the definition of ID in 2002 by the AAMR (Luckasson et al. 2002). The development of some instruments has been focused on the discriminatory power of the assessment instrument just around two SD units below the mean to make the diagnosis of ID more reliable (Tasse et al. 2012,

Balboni et al. 2014, Tasse et al. 2016). Besides being a part of the diagnostic assessment, measurement of adaptive abilities has been used when measuring both effectiveness of rehabilitation and evaluating decline with age (Sechoaro et al. 2014, Arvio and Luostarinen 2016). However, there is no unanimous agreement on the factors constituting adaptive abilities, or the best way to measure them. Neither is there clear understanding on the relationship between adaptive ability and intelligence (La Malfa et al. 2014, Price et al. 2018).

In addition to the behaviourally oriented scales, there has been an interest in developing more developmentally oriented approaches. One of them is The Scheme for Appraisal of Emotional Development, SAED (Dosen 2005a, 2005b), and the newer version, the Scale for Emotional Development-Revised, SED-R (Vandeveldt et al. 2016). Besides having a good correlation with one of the most widely used adaptive behaviour scales, The Vineland Social Maturity Scale (La Malfa 2014), it aims to give developmental insights. Emotional development and personality development are viewed as the developmental components that play important roles in adaptive and maladaptive behaviour as well as in the onset and presentation of psychopathology. A wider frame of mind is suggested, a replacement of the three-dimensional paradigm (bio–psycho–social) by a four-dimensional one (bio–psycho–socio–developmental) for the assessment and diagnosis of persons with ID.

2.2 RESEARCH METHODS IN THE EPIDEMIOLOGY OF ID

By 1969, the methods used in the epidemiology of ID could be roughly divided into three categories (Lemkau and Imre 1969):

1. Agencies likely to see cases of mental retardation were invited to report such cases.
2. A sample of households was visited, and relatives were asked about cases of ID by responding to key items on a questionnaire, and subsequently the rates were extrapolated for the total population.
3. Households were visited, suspected cases were ascertained and examined clinically, and rates were then calculated for the population.

In several countries persons with ID have been registered in national databases, such as found in Ireland (Kelly 2015), Western Australia (Pettersson et al. 2005, Bourke et al. 2018), and Taiwan (Lai et al. 2013). Register-based studies still form the majority in epidemiological research into ID. The first population sample studies were the so-called Finland-in-Miniature study in 1966 (Amnell 1966, Ruoppila 1966, Tarvainen 1966) and the Isle of Wight study in England (Rutter et al. 1970). The current literature review covers 83 studies, 75 from high-income countries and eight from low- and middle-income (LAMI) countries. Fifty (60%) of the studies are register-based, 29 (35%) are population sample studies, and four (5%) are household surveys. In the last group all were performed in LAMI countries. Strengths and limitations of various designs in epidemiological research into ID are listed in *Table 2*.

Population sample studies enable exact prevalence calculation, since the basic population is well-defined. They may be comprehensive if all individuals in the sample have been screened with sensitive methods and all the positives have been investigated. However, those constructing diagnostic classification systems (IAADD, ICD, DSM) acknowledge that it is clinical judgement which ultimately is important in diagnosis (Luckasson and Schalock 2015). It may take a longer time to make such a judgement in borderline cases than is possible in a cross-sectional study. The best method is long-term

follow-up, where the same individuals are assessed several times during their development. However, even in a longitudinal study the individuals may not have been assessed repeatedly for ID. In contrast, that may be the case before a person is noted in a register/database.

Table 2. Various types of study design and data collection methods in epidemiological research into ID, and their strengths and limitations.

Method	Explanation and study examples	Strengths	Limitations
Population sample studies	Population sample studies target a pre-defined group of people and assess the individuals once cross-sectionally or follow the cohort longitudinally and assess it at several points of time. The study population may be the total population in the area (Rutter et al. 1970, Birch et al. 1970) or a random sample, e.g. of communes in the country (Ruoppila 1966). If the cohort is large, the study often includes a screening stage to find possible cases (for example low school performance) to be examined more thoroughly.	The risk population is defined, and new cases may be detected in the follow-ups.	Systematic error in case-finding may over- or underestimate the prevalence. Needs large samples because ID is relatively rare.
Prospective regional birth cohort studies	All persons born during a predefined period in a defined region belong to the risk population. They are followed up prospectively, and clinical investigations are made at certain time points or information is collated from available information sources, such as registers (Rantakallio and von Wendt 1986).	The risk population is well-defined. Long-term follow-up may lessen the number of false negatives.	Laborious, drop-outs e.g. due to migration.
Longitudinal studies	In a longitudinal study the population is assessed several times during the follow-up period.	It is possible to notice changes in individuals' performance.	Laborious, drop-outs.
Cross-sectional studies	Cross-sectional studies collect data from a population, or a representative subset, at a specific point in time (period). The study population may be a representative age cohort, all pupils in a school district, other geographical sample, door-to-door interview, or sample from one or several registers (van Schroyen Lantman-de Valk et al. 2006). In cross-sectional studies of the prevalence of ID, usually the latest psychological test results (from a series of variable sizes) are considered if all subjects are not assessed in the study protocol. A cross-sectional study based on a service register may thus rely on longitudinal follow-up, which at some point has brought the individual into the register.	Relatively easy and inexpensive.	One time-point assessment often underestimates the prevalence. Risk of participation bias.
Screening	There may be a regular screen for developmental disorders at a certain age, or placement in special education based on previous evaluations. This screening may also be organized by the researchers with the methods explained (Rutter et al. 1970, Kääriäinen 1987). Screening may be done via registers and supplemented, e.g. by asking school teachers and nurses which individuals they think suffer from ID (Hagberg et al. 1981).	Helps to limit the number of persons in the risk population to be assessed.	Lack of sensitivity in the screening procedure leads to false negatives.
Register-based studies	Many kinds of registers are increasingly kept in social and health care, and education for the purposes of service delivery, and administering benefits and allowances (Van Naarden Braun et al. 2015). Sophisticated means can be developed for data mining (Lin et al. 2013). Persons can also be traced via contact with authorities, without a formal register (Gould 1976).	Information in registers is readily available.	For research purposes the criteria for registration may be compromised (problem of validity), or coverage not known (problem of reliability).
Intellectual Disability Databases	Information in several registers which are kept for social, health, and education purposes may be combined via linkage (Gissler et al. 1998, Bourke et al. 2018).	Combining may give a better coverage, when each register has some specific purpose, which does not concern all persons with ID.	Preventing double counts is a critical issue where there is not a unique personal/national identity code.
	In some countries systematic data is collected about persons with ID, concerning their characteristics, for the purposes of service delivery (Massey and McDermott 1996, Petterson et al. 2005, van Bakel et al. 2014). The information content in these databases may be very broad, and thus give good opportunity to carry out several types of studies on ID. However, the criteria for eligibility may be very varied and unsystematic.	The information is ready available.	Coverage is not complete, because it is usually restricted to those accepted in the services. The number of false negatives is not known.

2.3 PREVALENCE OF ID

2.3.1 High-income countries

The classification of countries in this review into high- or low- and middle-income (LAMI) economies is based on the World Bank's classification in June 2017 (World Bank 2017).

2.3.1.1 Children

In total, 56 studies concerning the prevalence of ID in childhood/adolescence in high-income countries are included in this review (*Table 3*, 58 articles, two studies being published in two separate papers, Gustavson et al. 1977b and Blomqvist et al. 1981, and Stein 1976a, 1976b). According to the study method, 31 are register-based and 25 are population sample studies. Of all, 79% were published before 2000 (see *Figure 1*). On average, population sample studies are newer than register-based studies.

According to study location, 36 studies were from Europe, 13 from North America, 3 from Asia, and 4 from Australia.

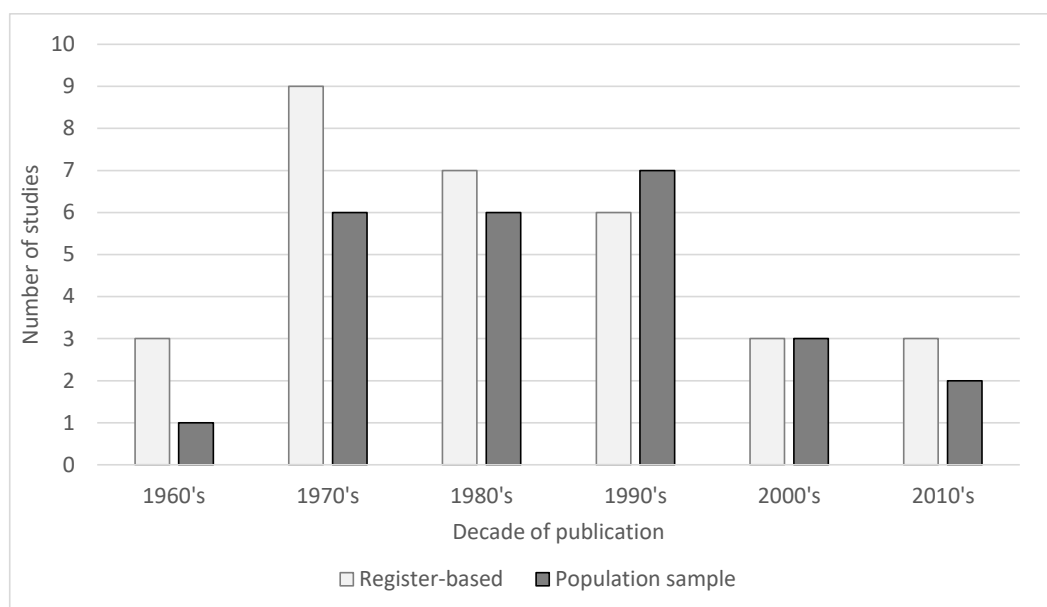


Figure 1. Studies of the prevalence of ID in childhood/adolescence in high-income countries according to study type and time of publication.

The prevalence of sID ($IQ \leq 50$) appeared to be similar across all reviewed studies, irrespective of study design (*Figure 2*). The median in this set was 0.41% (mean 0.44%, SD 0.14%, range 0.20% to 1.03%). Although the prevalence of sID did not seem to depend on the age of the study population, there may be some relationship between age and prevalence of sID. In most of the studies the prevalence was given as an average for an age span (see *Table 3*). This way of expressing the results may smooth possible differences in prevalence by age.

The prevalence of mID, in contrast, was more variable (range 0.15% to 4.59%). The median in this set was 0.54% (mean 0.92%, SD 1.05%). The prevalence of mID also did not seem to depend on age of the study population.

The proportion of mID reported was very variable (range 31% to 90%). The median in this set was 56% (mean 57%, SD 14%). Only in four studies was the proportion of mID above 80%. The population sample study by Birch et al. (1970) in the UK relied on a screen of the whole population under investigation, and the proportion of cases of mID was 86%. Likewise, the population sample study by Rutter et al. (1970) in the UK covered the whole study population on the Isle of Wight, and the proportion of cases of mID was 88%. In the population sample study by Broman et al. (1987) the proportion of mID for blacks was 87%, but 68% for whites. The highest proportion (90%) was noted in the register-based study by McDermott (1994) in South Carolina, USA. The definition of ID was based on the South Carolina State Department of Education Handbook, written for educational purposes. It is uncertain how well the definition matches those of AAMR/AAIDD.

Total prevalence was also very variable (range 0.35% to 5.50%). The median in this set was 1.12% (mean 1.45%, SD 1.18%). Total prevalence did not seem to depend on age of the study population when different studies were compared (*Figure 2* and *Figure 3*). However, an increase of prevalence by age was noted in studies where prevalence was expressed for different age groups (*Figure 5*).

It has been consistently noted that the prevalence of ID is higher among boys than girls (Stevens and Heber 1964, Dobbing et al. 1984). In a meta-analysis the prevalence was higher in males in both child/adolescent and adult populations (Maulik et al. 2011). Among adults the female-to-male ratio varied between 0.7 and 0.9, while in children/adolescents it varied between 0.4 and 1.0.

Table 3. Reviewed studies of sID and total ID in childhood and adolescence in high-income countries, in order of the year of publication. Population sample studies are shaded grey. The result is marked with asterisk () when there were several prevalence values/age spans available, and the maximum prevalence was chosen (almost always the oldest age span). Two studies on 19-year-olds are included, i.e. Granat and Granat 1973, and Stein et al. 1976a, 1976b.*

Study, Country	Sample, Design, N	Methods n = number of detected cases	Results Prevalence / age span	Remarks Coverage of levels. Showing in the Figures of this review
Goodman and Tizard 1962, UK, England, County of Middlesex	Register-based (local health records and further inquiries), N=451,800 (158,978 / 10–14 y)	Case finding: relying on existing records. Assessment methods used in clinical praxis not documented. n=1142 (574 / 10–14 y)	sID 0.36% / 10–14 y *	Only sID
Drillien et al. 1966, UK, Scotland, Edinburgh	Register-based (records from public health department + school medical service + additional inquiries), children born 1950–56, themselves and/or mother resident in Edinburgh in 1962–64, N=39,498	Case finding relied on registration. Ascertainment at the time of registration, at least one test available, IQ<70, n=446	1.13% / 7.5–14.5 y sID 0.50% / 7.5–14.5 y	Proportion of mID 56%
Ruoppila 1966, Finland	Population sample (representative stratified random sample of whole Finland, by geographical regions, "Finland-in-Miniature" study in 1962), N=35,560 (15–19 y)	Case finding from various sources, suspected cases screened by a selection of psychological tests, psychological assessment for positively screened, AAMR criteria (Heber 1961), IQ <70, n=335 (15–19 y). Predictive validity assessment of ID diagnoses 7 years later Ruoppila (2011). Stanford–Binet Scale, IQ <52, n=14	0.94% / 15–19 y * sID 0.50% / 15–19 y *	Proportion of mID 47%. Displayed also in line graph <i>Figure 5</i> .
Åkesson 1967, Sweden, Islands	Register-based (local registers + inquiries from various officials), N=2525		sID 0.55% / 10–20 y *	Only sID
Birch et al. 1970, UK, Scotland, Aberdeen	Population sample (geographically defined area), N=8274	Psychometric tests, IQ <75, n=227	2.74% / 8–10 y sID 0.37% / 8–10 y	High cut-off: Proportion of mID 86%
Rutter et al. 1970, UK, England, Isle of Wight	Population sample (geographically defined area), N=2334	School screening + IQ test, n=59	2.52% / 9–11 y sID 0.30% / 9–11 y	Test was normalized in the study population. Proportion of mID 88%
MacKay 1971, UK, Northern Ireland	Register-based (registers of three regional special care management committees by the end of 1968), N=574,600 (0–19 y)	Ascertained to need special care, IQ <50, n=594 (15–19 y)	sID 0.47% / 15–19 y *	Only sID.
Brask 1972, Denmark, County of Aarhus	Register-based (local register, the Mental Retardation Service concerning children from the county of Aarhus), N=52,880 (0–14 y), 18,453 (10–14 y)	Ascertainment before registration, methods not explained, n=155 (10–14 y)	0.84% / 10–14 y * sID 0.37% / 10–14 y *	Proportion of mID 56%. Displayed also in line graph <i>Figure 5</i> .
Granat and Granat 1973, Sweden	Population sample (examination for military service, 19-year-olds), N=1,872,609.	A validated test given to all registering men, IQ <70.	1.50% + 0.71% previously classified as having ID = 2.21% / 19 y	Not included in bar graph <i>Figure 2</i> due to age > 18 y. Discussed on page 45.
McDonald 1973, Canada, Quebec	Population sample (cohort study, children born in 1958 and living in Quebec 1966–69), N=141,500.	Test of mental development, Quick Screening Scale, n=507.	sID 0.38% / 8–12 y	Only sID.

Table 3. Reviewed studies of prevalence of sID and total ID in childhood and adolescence in high income countries, continued ...

Study, Country	Sample, Design, N	Methods n = number of detected cases	Results Prevalence / age span	Remarks Coverage of levels. Showing in the Figures of this review
Amnell 1974, Finland, Helsinki	Population sample (birth cohort follow-up until 14 y, all children born in 1955 in Helsinki), N=6,408.	Psychological tests were checked from medical records, IQ 0–67, n=56	0.91% / 14 y sID 0.39% / 14 y	Proportion of mID (IQ 52–67) 57%
Bernsen 1976, Denmark	Register-based (national and regional registers), N=50,667 (0–14 y)	Psychological tests were evaluated from previous registered records, re-test if IQ 50–60, inclusion when IQ <50, n=66(n=155 (10–14 y))	sID 0.45% / 10–14 y *	Only sID
Gould 1976, UK, England, London area	Register-based (local authorities), N=34,900	Psychological test, IQ <50, n=150.	sID 0.43% / 0–14 y	Only sID
Reynolds 1976, Australia, Queensland	Register-based, (various local authors), N=396,200	Psychological tests, IQ <55, n=1350	sID 0.34% / 5–16 y	Only sID
Stein et al. 1976a, 1976b Netherlands	Population sample (examination for military service for 19-year-old males born in 1944–1947), N=405,548	Three criteria used in combination: history of special schooling, Raven intelligence test score, and clinical ICD diagnosis.	3.0%–6.1% / 19 y sID 0.37% / 19 y	Not included in bar graph <i>Figure 2</i> due to age > 18 y. Discussed on page 49.
Frost 1977, Western Ireland	Register-based (regional mental handicap assessment centre, children born 1956–1970), N=60,364.	Psychological tests, n=578	0.96% / 4–19 y sID 0.38% / 4–19 y	Proportion of mID 60%
Gustavson et al. 1977a, Sweden, County of Uppsala	Register-based, (national register + services), N=16,779	Psychological tests IQ <50, n=55	sID 0.33% / 11–16 y	Only sID.
Gustavson et al. 1977b, Sweden, County of Västerbotten	Register-based, (national register for service provision), N=40,871 (5–16 y)	Psychological tests, IQ <50, n=74(13–16 y)	sID 0.53% / 13–16 y *	Only sID. Combined with Blomqvist et al. 1981 in <i>Figure 2</i> .
Laxova et al. 1977, UK, England, Hertfordshire	Register-based (regional authorities), N=46,960.	Psychological assessment, IQ≤50, n=146	sID 0.31% / 7–9 y	Only sID.
Blomqvist et al. 1981, Sweden, County of Västerbotten	Register-based, (national register for service provision), N=40,871 (8–19 y)	Psychological tests, IQ=50–69, n=75 (16–19 y)	mID 0.47% / 16–19 y *	Only mID. Includes Gustavson et al. 1977b in <i>Figure 2</i> .
Elliot et al. 1981, UK, England, Oxfordshire	Register-based, (local authorities), N=81,401	Not described, assessment at the time of inclusion to services, n=337	sID 0.41% / 5–14 y	Only sID.
Lindsey and Russel 1981, UK, England, Cornwall	Register-based (local authorities), N=82,500 (5–18 y)	Psychological tests, n=123 (15–18 y).	sID 0.47% / 15–18 y *	Only sID.
Mitchell and Woodthorpe 1981, UK, England, London	Register-based (local authorities), N=56,140	Psychological tests, IQ <50, n=282	sID 0.50% / 15–20 y	Only sID, includes a minority of persons with milder ID but equal use of services.
Fishbach and Hull 1982, Canada, province of Manitoba	Register-based (several hundred major agencies were contacted), N=967,042 (all ages)	Ascertainment based on register information: psychometric assessment (55%), most recent medical assessment (39%), opinion of primary caregiver or educational specialist (15%).	1.26% / 12–17 y *	Levels of ID not available by age groups. Prevalence given for non-institutionalized persons. Displayed also in line graph <i>Figure 5</i> .

Table 3. Reviewed studies of prevalence of sID and total ID in childhood and adolescence in high income countries, continued ...

Study, Country	Sample, Design, N	Methods n = number of detected cases	Results Prevalence / age span	Remarks Coverage of levels. Showing in the Figures of this review
Gillberg et al. 1983, Sweden, Gothenburg	Population sample (children born in 1971 and living in Gothenburg, Sweden, by the end of 1977), N=5114.	Preschool questionnaire screening (for diverse neuropsychiatric problems) + IQ test (WISC performance), n=47	0.91% / 8 y sID 0.35% / 8 y	Screening instrument's sensitivity to detect persons with ID not known. In addition to persons with ID, almost 1% with borderline IQ.
Richardson et al. 1984, UK, England, one city (unnamed)	Register-based, children born 1955–1955, N=13,842.(5–22 y)	Placement in ID services before 15 y of age, no psychometric definition was used, n=206 (11 y)	1.47% / 11 y *	Almost three quarters of those who had been at school for educable, mentally retarded children received no services in the young adult period.
Shiotsuki 1984, Japan, Kurume City	Population sample (all children born in 1969–1974), N=21,622	All children screened, IQ test results collected from previous records, IQ < 70, n=154.	0.71% / 7–12 y sID 0.49% / 7–12 y	Proportion of mID 31%.
Rantakallio and von Wendt 1986, Finland, two northernmost provinces	Population sample (geographically defined birth cohort formed prenatally, follow-up 14 y), N=11,766.	Screening: sub-average performance at school. Ascertainment: psychological test results for IQ were collected from different sources, n=140.	1.19% / 14 y sID 0.63% / 14 y	Proportion of mID 47%.
Broman et al. 1987, USA, various regions	Population sample (women enrolled during pregnancy into the collaborative Perinatal Project in 12 urban medical centres, followed until 7 y of age), N=36,851.	Psychological assessment (abbreviated version of the Wechsler Intelligence Scale for Children (WISC, IQ <70) to all subjects, clinical judgement, n=1317	Whites 1.68% / 7 y Whites sID 0.53% / 7 y Blacks 5.27% / 7 y Blacks sID 0.68% / 7 y	48% white, 52% black, proportion of mID 68% for whites, and 87% for blacks. In bar graph <i>Figure 2</i> estimates for whites and blacks are shown separately. Discussion on page 45.
Hagberg et al. 1987, Sweden, Gothenburg	Population sample (an unselected series of children residing in Gothenburg, Sweden, and born between 1966–70), N=23,544	Information traced from multiple registries. IQ tested with the Swedish standardization of the WISC or the Terman–Merrill test before registration, n=170	0.72% / 14–18 y sID 0.33% / 14–18 y	Proportion of mID 54%
Kääriäinen 1987, Finland, district of Kuopio	Population sample (both cohorts 1969–1970 and 1971–1972, assessed at the age of 8–9 y), N=6797+6085.	Screening by school tests, individual IQ tests, n=60+118.	1.38% / 8–9 y sID 0.63% / 8–9 y	Larger difference between two screened populations than 95% CI would assume. IQ test was normalized in the study population.
McQueen et al. 1987, Canada, Maritime region	Register-based (school records + agencies), N=84,109.	Psychological test, sID IQ <56, n=307	sID 0.37% / 7–10 y	Only sID
Díaz-Fernández 1988, Spain	Register-based (regional register), N=2,754,700 (all ages)	IQ tests when registered which may happen when applying for health care or social benefits, n=1632 (15–19 y)	0.91% / 15–19 y * sID 0.49% / 15–19 y *	Proportion of mID 31% in total sample. Displayed also in line graph <i>Figure 5</i> .
Andersen et al. 1990, Denmark, Fredriksborg county	Population sample (complete county birth cohort surveyed at age 4), N=4138	Case-finding by 108 general practitioners plus checking from available registers. Psychological assessment and clinical judgement, n=176	0.43 % / 4 y sID 0.29% / 4 y	Proportion of mID 33% (young age group).
Benassi et al. 1990, Italy, Bologna	Register-based (school + services registers), N=26,494	Psychological test, n=90	sID 0.34% / 6–13	Only sID.

Table 3. Reviewed studies of prevalence of sID and total ID in childhood and adolescence in high income countries, continued ...

Study, Country	Sample, Design, N	Methods n = number of detected cases	Results Prevalence / age span	Remarks Coverage of levels. Showing in the Figures of this review
Cooper 1990, Germany	Register-based (local authorities), N=35,026	Psychological test, n=245.	0.70% / 7–16 y sID 0.37% / 7–16 y	Proportion of mID 47%
Wellesley et al. 1991, Western Australia	Population sample (all children born 1967–76), N=210,789	Registers contained psychometric test results. IQ <70, n=1602	0.76% / 6.5 y sID 0.36% / 6.5 y	Proportion of mID 38%
Yeargin-Allsopp et al. 1992, USA, Georgia, Atlanta	Register-based (multiple sources, children born 1975–1977), population size per year N= ca. 34,000.	Registers contained psychometric test results. Inclusion only by IQ ≤70, n=1074	1.03% / 10 y	No information of level of ID
McDermott 1994, USA, South Carolina	Register-based (children enrolled for mental handicap programmes during school year 1980–81 in 92 independent school districts), N=616,000.	ID definition for educational purposes according to the South Carolina State Department of Education Handbook, n=25,691.	4.17% / 5–14 y	Different school districts classified pupils very differently, including variably slow learners and learning-disabled pupils. No information on level of ID.
Murphy et al. 1995, USA, Georgia, Atlanta metropolitan area	Population sample (children born 1.1.1975–31.12.1977, and residing in the study area at age 10 y, records reviewed from the public schools (98%) or other sources), N= 89,534.	Inclusion with previous IQ tests. IQ ≤70, n=1,074.	Whites 0.47% / 10 y Whites sID 0.27% / 10 y Blacks 1.45% / 10 y Blacks sID 0.52% / 10 y	Proportion of mID for whites 43%, for blacks 64%. In bar graph <i>Figure 2</i> estimates for whites and blacks are shown separately.
Fernell 1996, Sweden	Population sample (children aged 9–15 years, born in 1979–85 and living in Botkyrka, one of the 24 suburban municipalities in Stockholm, on December 31, 1994), N=6397	Children were traced from schools and hospitals. Psychological test for all. IQ 50–72, n=82	mID 1.28% / 9–15 y	Only mID. 34% of the inhabitants had their roots in foreign countries (foreign nationals or born in a foreign country). The number of children with at least one parent born in a foreign country was about 50%. Cut-off for IQ higher than normal.
Katusic et al. 1996, USA, Montana	Population sample (birth cohort of children born from 1976 to 1980 inclusive), N=3287	Follow-up through school and community medical records for diagnostic criteria of mild or severe mental retardation, n=30	0.91% / 8 y sID 0.49% / 8 y	Proportion of mID 47%.
Massey and McDermott 1996, USA, All states	Register-based (U.S. Department of Education, children who were enrolled in special education programmes), population data not given.	National guidelines determine the eligibility requirements for entitlement programmes, administered locally and subject to local interpretations and modifications. Only calculated prevalence rates given.	1.14% / 6–17 y	Large differences between states, see <i>Figure 6</i> .
Camp et al. 1998, USA, different regions	Population sample (women enrolled during pregnancy into the collaborative Perinatal Project in 12 urban medical centres, followed until 7 y of age), N=35,684.	Psychological assessment (abbreviated version of the WISC to all subjects), clinical judgement. IQ <70, n=1311.	Whites 1.7% / 7 y Blacks 5.5% / 7 y	48% white (lower SES 33.2% of them), 52% black (lower SES 64.7% of them). Levels of ID not given. Proportion of mID not known. In bar graph Discussion on page 45.

Table 3. Reviewed studies of prevalence of sID and total ID in childhood and adolescence in high income countries, continued ...

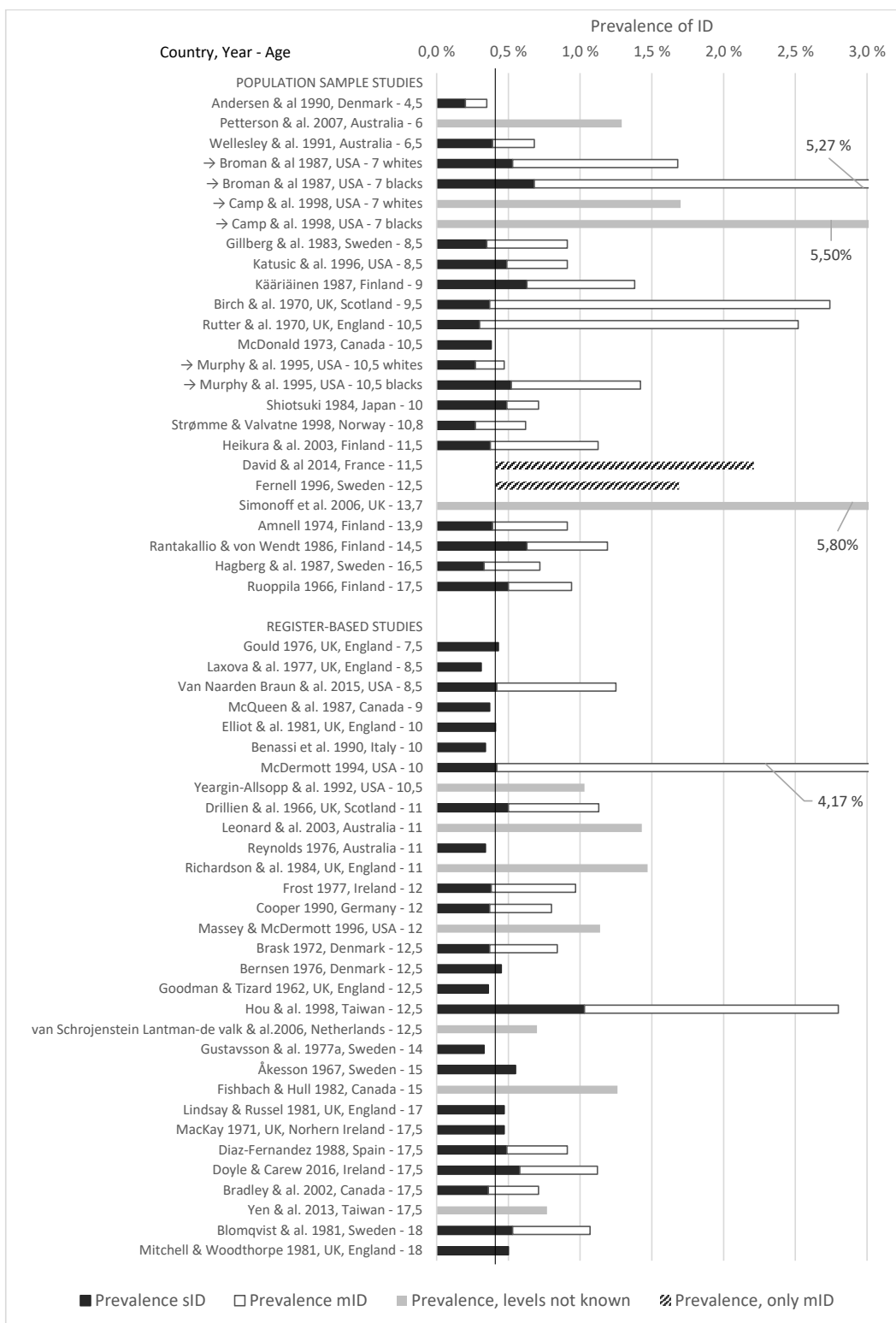
Study, Country	Sample, Design, N	Methods n = number of detected cases	Results Prevalence / age span	Remarks Coverage of levels. Showing in the Figures of this review
Hou et al. 1998, Taiwan, Taipei	Register-based (subjects enrolled from the authors' outpatient clinic, and all schools or institutions in study area), N=423,000.	Case finding: no details given of the "nationwide screening program". Ascertainment: no details given of the diagnostic assessment. IQ ≤ 70 , n=11,892	2.81% / 6–18 y sID 1.04% / 6–18 y	Recruitment less than 10% of total population, selection bias not known. Proportion of mID 63%
Stromme and Valvatne 1998, Norway, County of Akershus	Population sample (all children born 1980–1985) N=30,037	Cases ascertained from multiple sources and psychometrically assessed, DSM-IV, IQ ≤ 70 , n=185	0.62% / 10.8 y sID 0.27% / 10.8 y	Proportion of mID 56%
Bradley et al. 2002, Canada, Ontario	Register-based (schools, services, Intensive case finding), N=35,485	Psychological test, IQ ≤ 75 , n=255	0.72% / 14–20 y sID 0.35% / 14–20 y	Proportion of mID 49%
Heikura et al. 2003, Finland, Two northernmost districts	Population sample (birth cohort 1985–86, follow-up to 11.5 y), N=9351.	Screening: sub-average performance at school. Ascertainment: psychological test results for IQ were collected from different sources, n=105.	1.12% / 11.5 y sID 0.37% / 11.5 y	Proportion of mID 67%
Leonard et al. 2003, Western Australia	Register-based (Disability Services Commission, and three agencies providing educational support for Western Australian children with ID)	Ascertainment: (1) IQ < 70 on formal testing; (2) a condition known to be associated with intellectual disability (e.g. Down's syndrome); or (3) clearly documented as having intellectual disability in Disability Services Commission records.	Average prevalence 6–15 y 1.43%	Prevalence rate exceptionally decreasing by age. Levels are given as mild+moderate vs. severe+profound. Proportion of mild+moderate 74%. Displayed also in line graph Figure 5.
Simonoff et al. 2006, UK, London	Population sample (secondary schools in the London Borough of Croydon), N=8841.	Screening with The Cognitive Abilities Test, in-depth psychometric assessment was performed in a sample with the WISC-III test battery. Prevalence estimated using different imputation methods and assumptions about individuals not screened.	5.8% to 10.6% / 13.7 y y 1.43%	Participation rates were low. Children with severe and profound learning disabilities not included in the original sampling frame. Nevertheless, the prevalence of IQ ≤ 60 was ca. 2.5%. Discussed on pages 41 and 47.
van Schroyensteen Lantman-de Valk et al. 2006, Netherlands, Province of Limburg	Register-based (combination of general practice databases and service registrations), N=261,700 (0–19 y)	AAMR 1992 or modified criteria based on service needs, n=1511 (5–19 y)	0.70% / 5–19 y *	No method available to estimate the size of not being among the services. Proportion of mID not known. Displayed also in line graph Figure 5.
Pettersen et al. 2007, Western Australia	Population sample (all children born in area 1980–1999 and surviving to 1 y, case identification from multiple sources into the IDEAS database), N=474,285.	Inclusion criteria IQ < 70 before the age of 18 y, may include cases where either verbal or performance IQ falls within borderline range (70–84), n=6106.	1.29% / 1–10 y	Levels given as mild+moderate vs. severe+profound. Proportion of mild+moderate 90%.
Yen et al. 2013, Taiwan	Register-based (National Disability Register), N=5,849,303 (0–19 y)	Inclusion criteria in the register not described, n=30,918	0.77% / 15–19 y	Proportion of mID not known. Displayed also in line graph Figure 5.
David et al. 2014, France	Population sample (children born in 1997 residing in Isère County, France, in 2008), N=15,100	Psychometric test results collected from administrative records, IQ 50–69, n=267	1.8% / 9–13 y	Only mID

Table 3. Reviewed studies of prevalence of sID and total ID in childhood and adolescence in high income countries, continued ...

Study, Country	Sample, Design, N	Methods n = number of detected cases	Results Prevalence / age span	Remarks Coverage of levels. Showing in the Figures of this review
Van Naarden Braun et al. 2015, USA, metropolitan Atlanta	Register-based (developmental disabilities surveillance program for 8-year-olds in metropolitan Atlanta, 1991–2010), N=not available.	Systematic review of developmental evaluations at multiple education and health sources, IQ ≤ 70 on the most recently administered test of intellectual ability, n=5590	1.30% / 8 y sID 0.42% / 8 y	Average prevalence across the period 1991–2010. Size of the general population based on a combination of several censuses.
Doyle and Carew 2016, Ireland	Register-based (national database for service provision, intended to cover all with sID, and others who need special services for ID), N=28,108 (all ages)	Individuals registered in the database via a multidisciplinary assessment process, n=3175 (15–19 y)	1.12% / 15–19 y * sID 0.58% / 15–19 y	Proportion of mID 45% (15–19 y). Displayed also in line graph <i>Figure 5</i> .
Maenner et al. 2016, USA, All 50 states	Population sample (multistage sampling). Two different national surveys, NSCH 2011–2012 N=85,637, NHIS 2011–2013 N=34,503	Random digit-dial telephone survey for parental view if ID has ever been suspected.	NSCH: 1.22% / 2–17 y, 1.5% / 14–17 y * NHIS: 1.21% / 2–17 y, 1.78% / 10–13 y *	Response rate low, 23% in 2011–2012, 69–75% in 2011–2013. No verification of parental information. Not included in bar graph <i>Figure 2</i> due to lack of verification.

SES = socioeconomic status, NSCH = National Survey of Children's Health, NHIS = National Health Interview Survey

Figure 2. (Next page) Prevalence of ID in childhood in high-income countries; studies ordered by increasing age of study population. From each study the oldest age group with highest prevalence is selected and the prevalence is plotted at the average age of the age span given. Blomqvist et al. 1981 include an estimate for sID from the same data as Gustavson et al. (1977b). Fernell (1966) and David et al. (2014) gave only mID, which is plotted above the median value of sID. Where estimates for white and black populations are given separately, there is an arrow in front of the study author. Vertical line shows the median sID (0.41%).



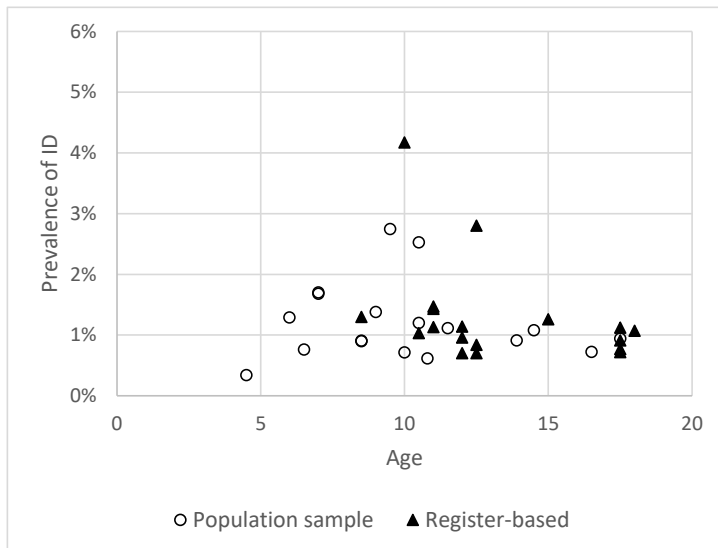


Figure 3. Prevalence of ID by age in all reviewed studies providing this information, according to study type; 18 population sample studies and 18 register-based studies. Only data concerning white population is included from studies by Broman et al. 1987, Camp et al. 1988, and Murphy et al. 1995. From each study the oldest age group with highest prevalence is selected and the prevalence is plotted at the average age of the age span given.

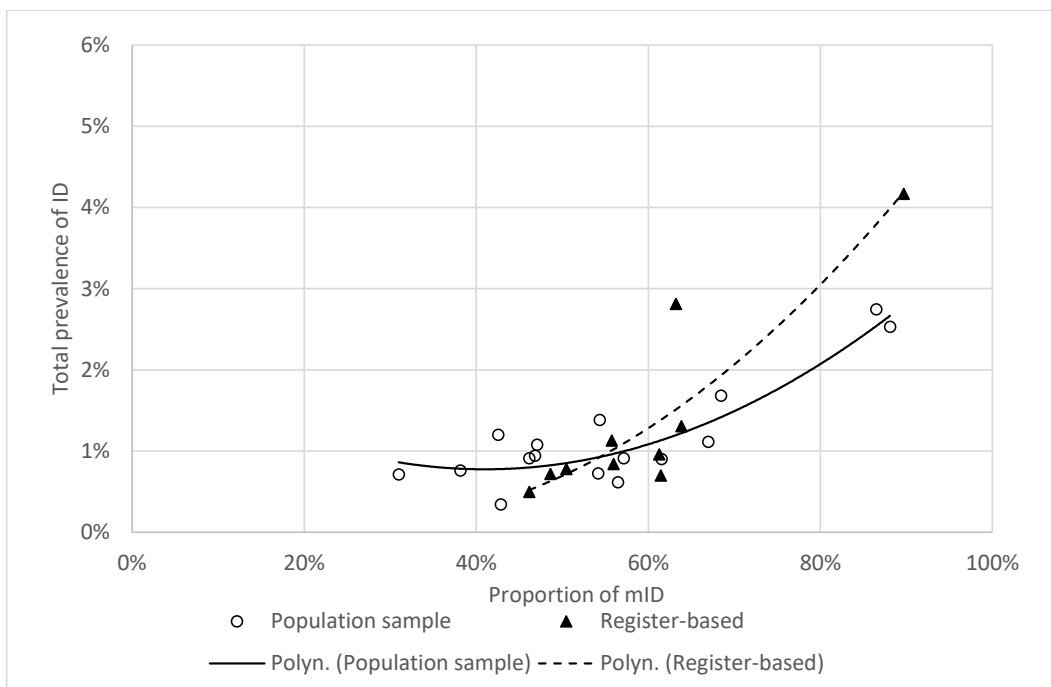


Figure 4. Total prevalence of ID by proportion of mID in all reviewed studies providing this information, according to study type; 16 population sample and 11 register-based studies. Only data concerning white population is included from studies by Broman et al. 1987 and Murphy et al. 1995. For each set a second-order polynomial trend line is added¹.

¹ A second order polynomial trend line was chosen as a linear one was not possible. The trend line was drawn by using an MS Excel spreadsheet.

Estimates of total prevalence are of course dependent largely upon the inclusion of persons with mID in the study sample, while the prevalence of sID is relatively constant. In *Figure 4* total prevalence estimates are plotted against the reported proportion of mID for all included studies where both mID and total prevalence were given.

Eight studies where a prevalence estimate was given for different age groups in childhood and adolescence are reproduced in *Table 4*, with additional details of prevalence by age groups, and the data are plotted in *Figure 5*. All but one shows an increase of prevalence by age. The one showing the opposite trend (Leonard et al. 2003) was a study of a mixed population of Caucasian and Aboriginal children. The prevalence rate was connected to ethnic background; 1.32% for Caucasian vs. 3.08% for Aboriginal, which explains the high estimate. The reason for decreasing prevalence by age is not discussed in the paper. A ninth study, not included in the table/figure due to its abnormally high prevalence rate, was also from Australia (Bourke et al. 2016). The prevalence rate for ages 5–18 increased from 0.98% to 2.42%. The study population was also mixed Caucasian/Aboriginal. The prevalence across all ages was 1.57% for Caucasians and 3.90% for Aboriginals. The two Australian studies relied on two data sources, the Disability Services Commission, and the Education Department. The difference by ethnic background was more pronounced in the educational source of ascertainment.

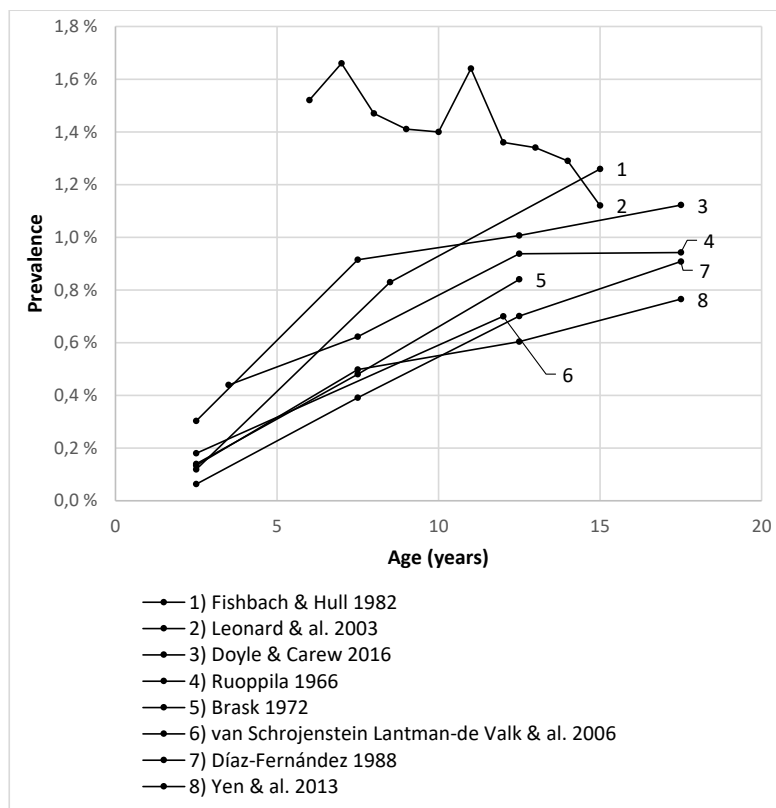


Figure 5. Prevalence of ID in childhood at different ages in eight studies. Prevalence values are plotted at the average age of each age span given in the study.

Table 4. Studies exploring the prevalence of ID in different age groups in childhood and adolescence in high-income countries, in order of the year of publication. Population sample study is shaded grey.

Study, Country	Sample, Design, N	Methods	Results Prevalence / age span	Remarks
Ruoppila 1966, Finland	Population sample (representative stratified random sample of the whole of Finland, "Finland-in-Miniature" study), N=147,240 (2–19 y)	Thorough psychological assessment, AAMR criteria (Heber 1961), IQ <70, n=1145 (2–19 y).	0.44% / 2–4 y 0.62% / 5–9 y 0.94% / 10–14 y 0.94% / 15–19 y	Proportion of mLD 45%
Brask 1972, Denmark, County of Aarhus	Register-based (local register, the Mental Retardation Service concerning children from the county of Aarhus aged 0–14 on census day, 31.12.1962). N=52,880.	Ascertainment before registration, methods not explained, n=260 (0–14 y)	0.14% / 0–4 y 0.48% / 5–9 y 0.84% / 10–14 y	Proportion of mLD 48%
Fishbach and Hull 1982, Canada, Province of Manitoba	Register-based (several hundred major agencies were contacted), N=967,042	Ascertainment based on register information: psychometric assessment (55%), most recent medical assessment (39%), opinion of primary caregiver or educational specialist (15%).	0.12% / 0–4 y 0.82% / 5–11 y 1.26% / 12–17 y	Proportion of mLD 36%. Prevalence given for non-institutionalized persons only. 90% agreed to participate.
Díaz-Fernandez 1988, Spain,	Register-based (regional register), N=2,754,700 (all ages)	IQ tests when registered which may happen when applying for health care or social benefits, n=13,636 (0–19 y)	0.06% / 0–4 y 0.39% / 5–9 y 0.70% / 10–14 y 0.91% / 15–19 y	Proportion of mLD 20–40%
Leonard et al. 2003, Western Australia	Register-based (Disability Services Commission, and three agencies providing educational support for Western Australian children with ID), N=240,358 (6–15 y)	Ascertainment: (1) IQ <70 on formal testing; (2) a condition known to be associated with intellectual disability (e.g. Down's syndrome); or (3) clearly documented as having intellectual disability in Disability Services Commission records, n=3426 (6–15 y)	1.52% / 6 y 1.66% / 7 y 1.47% / 8 y 1.41% / 9 y 1.40% / 10 y 1.12% / 15 y	Prevalence rate exceptionally decreasing by age. Levels are given as mild+moderate vs. severe+profound. Proportion of mild+moderate 74%.
van Schroyensteen Lantman-de Valk et al. 2006, Netherlands, Province of Limburg	Register-based (combination of general practice data bases and service registrations), N=261,700 (0–19 y)	AAMR 1992 or modified criteria based on service needs, n=1511 (0–19 y).	0.18% / 0–4 y 0.70% / 5–19 y	No method available to estimate the size of not being among the services
Yen et al. 2013, Taiwan	Register-based (National Disability Register), N=5,849,303 (0–19 y)	Inclusion criteria in the register are not described, n=30,918 (0–19 y)	0.13% / 0–4 y 0.50% / 5–9 y 0.60% / 10–14 y 0.77% / 15–19 y	Proportion of mLD not known.
Doyle and Carew 2016, Ireland	Register-based (national database for service provision, intended to cover all with sLD, and others who need special services for ID, N=1,262,609 (0–19 y)	Individuals registered in the database via a multidisciplinary assessment process, n=10,236 (0–19 y)	0.30% / 0–4 y 0.92% / 5–9 y 1.01% / 10–14 y 1.12% / 15–19 y	Proportion of mLD 36%

When estimating the prevalence of ID during the developmental period, it seems important to consider the age at which the estimate is based, because of the gradual increase of cumulative prevalence. As noted above, the proportion of mID also has a great influence on the estimate. In none of these studies was the proportion of mID above 50%, in two it was 45–48% (Ruoppila 1966, Brask 1972), in three it was less than 40% (Fishbach and Hull 1982, Díaz-Fernández 1988, Doyle and Carew 2016), and in three the information was not collected (Leonard et al. 2003, van Schrojenstein Lantman-de Valk et al. 2006, Yen et al. 2013).

2.3.1.2 *Adults*

Compared with studies on children and adolescents, there are far fewer ID prevalence studies on adult populations. In six studies (see *Table 5*) prevalence estimates were given for more than three different age groups (see *Figure 7*). On average the prevalence rates decline with age from an initial value of ca. 0.70–0.90% at a little above 20 years of age. At the age of 70–80 years, the prevalence has dropped to ca. 0.10–0.25%. However, the rate of decline varies greatly – at 50 years the range of estimates is ca. 0.30–0.75% (see *Figure 7*).

In the study by Massey and McDermott (1996) prevalence rates were collected in a national survey from different states of the USA, with same methodology. The prevalence rates for adults were 0.4%–0.7% in most of the eastern and western states, but on a remarkably higher level, 1.0%–1.6% in most of the east south-central and west south-central states.

Table 5. Studies on prevalence of ID in different age groups in adulthood in high-income countries, in order of the year of publication. Population sample study is shaded grey.

Study	Material	Methods	Results		Remarks
Ruoppila 1966, Finland	Population sample (representative stratified random sample of the whole of Finland; so-called Finland-in-Miniature study), N=194,000 (20–59 y)	Psychological assessment, AAMR criteria (Heber 1961), IQ<70, n=1306 (20–59 y)	0.91% / 20–24 0.60% / 25–29 0.61% / 30–34 0.61% / 35–39	0.65% / 40–44 0.67% / 45–49 0.65% / 50–59 0.78% / 55–59	Proportion of mID 42–50%, average 47%
Beange and Taplin 1996, Sydney, Australia	Register-based (cases were traced from various agencies and professionals). N=104,584 (20–50 y)	AAMR 1983, IQ ≤70. Review of previous psychological assessments within past 5 years; if not available new assessment performed, n=346 (20–50 y)	0.37% / 20–24 0.41% / 25–29 0.41% / 30–34	0.29% / 35–39 0.22% / 40–44 0.25% / 45–49	Proportion of mID 34%
van Schrojenstein Lantman-de Walk et al. 2006, Netherlands, province of Limburg	Register-based (service providers, General Practice Health Information System), N=889,000 (≥20 y)	AAMR 1992, IQ <70–75, n=5829 (≥20 y)	0.66% / 20–34 0.85% / 35–49 0.63% / 50–69 0.26% / 70+		Proportion of mID not available
McConkey et al. 2006, Ireland and UK, Northern Ireland	Register-based (National database for recipients of services in 2002). N=2,776,587 (Ireland), N=1'185'114 (UK, N Ireland) (≥20 y)	Psychological tests before entering the services, n=16,794 (Ireland), n=8340 (UK, N. Ireland), (≥20 y)	Ireland 0.93% / 20–34 0.82% / 35–49 0.67% / 50–64 0.22% / 65+	UK, N. Ireland 0.77% / 20–34 0.72% / 35–49 0.49% / 50–64 0.20% / 65+	Proportion of mID 30%
Dupont 1989, Denmark	Register-based (national service register for persons with ID, 1974). N=2,545,053 (≥21 y)	Psychological tests before entering the services, n=12,442 (≥21 y)	0.69% / 21–25 0.63% / 26–30 0.48% / 31–35 0.44% / 36–40 0.41% / 41–45 0.36% / 46–50	0.35% / 51–55 0.34% / 56–60 0.29% / 61–65 0.23% / 66–70 0.22% / 71–75 0.14% / 76+	Proportion of mID not available. Many people with mID leave the services after school.
Yen et al. 2013, Taiwan	Register-based (National Disability Register). N=16,912,101 (≥25 y)	Inclusion criteria in the register not described, n=64,470 (≥25 y)	0.59% / 25–29 0.47% / 30–34 0.39% / 35–39 0.34% / 40–44 0.30% / 45–49 0.26% / 50–54 0.25% / 55–59	0.23% / 60–64 0.16% / 65–69 0.12% / 70–74 0.10% / 75–79 0.09% / 80–84 0.12% / 85–89 0.28% / 90–94	Proportion of mID not available

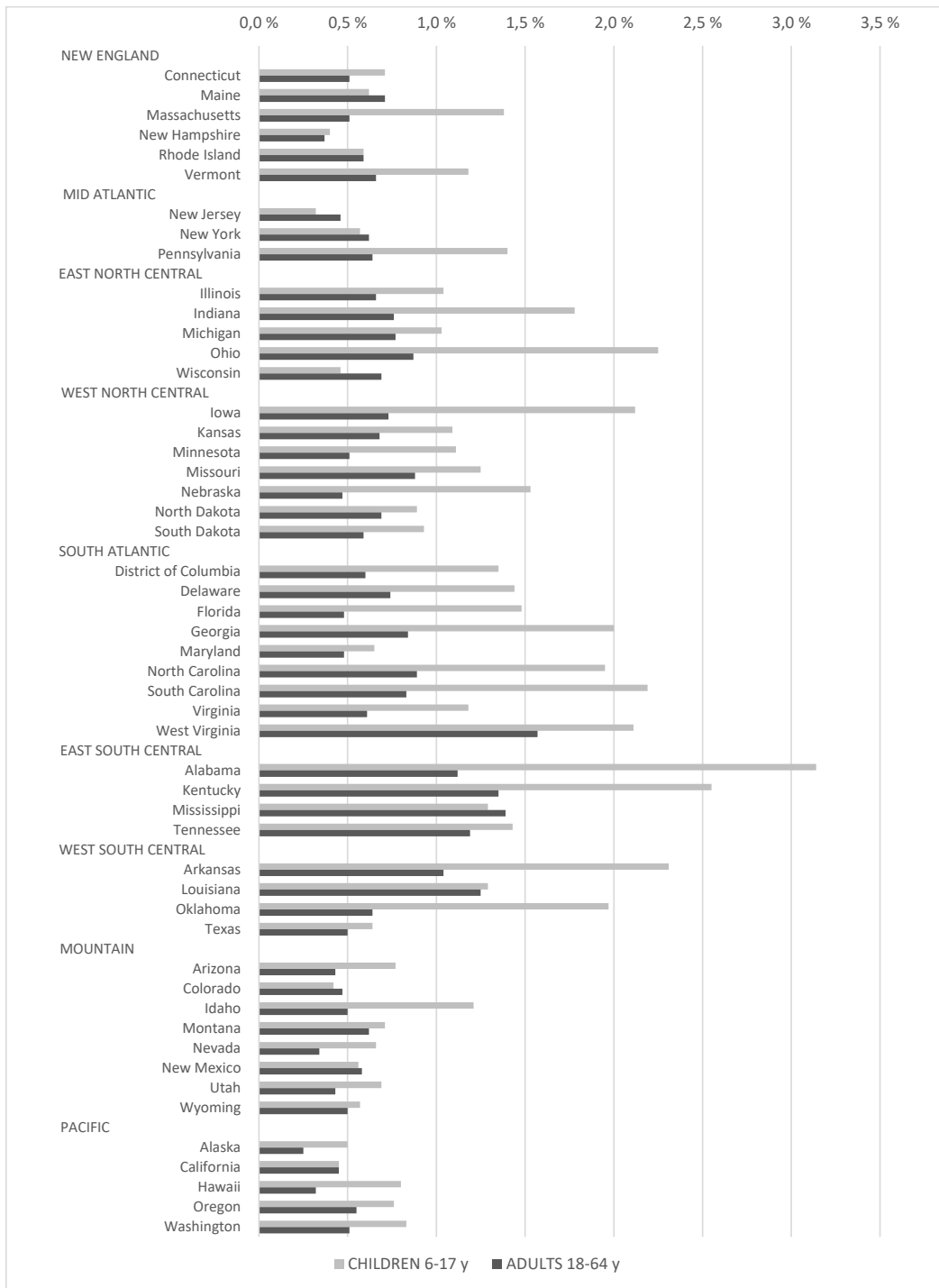


Figure 6. Prevalence of ID among children and adults in different states in the USA (according to Massey and McDermott 1996).

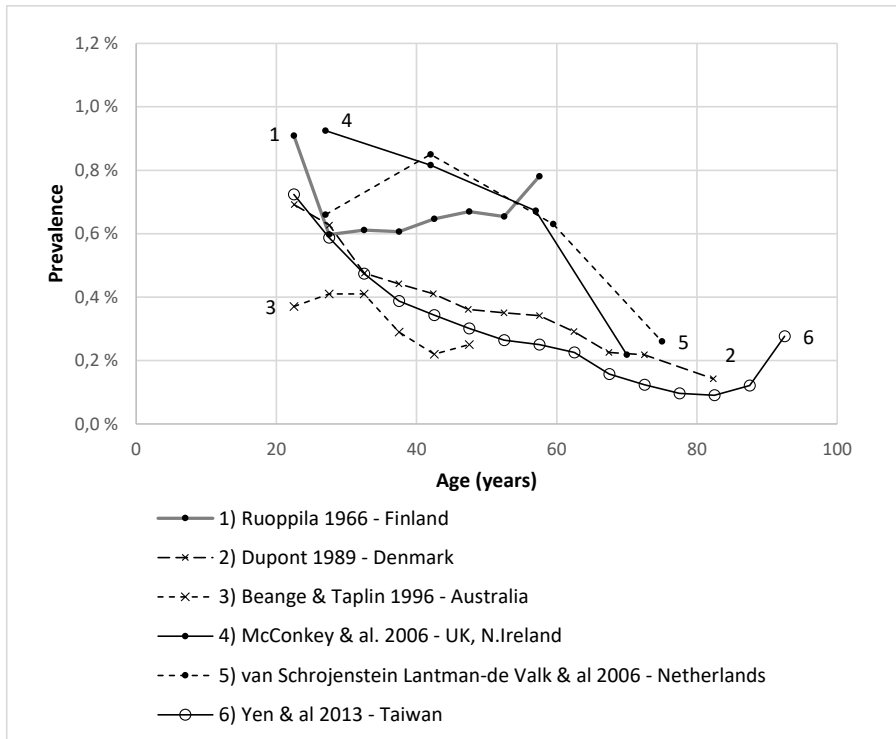


Figure 7. Prevalence of ID in adulthood at different ages in six studies.

2.3.1.3 Aging with ID

The epidemiology of ID among elderly people has been studied using various methods. *Table 6* summarizes the findings of fifteen studies. Persons with ID have a shorter life span than those in the general population, and the decrease in longevity is associated with the level of ID (Bittles et al. 2002, McCarron et al. 2015). In sID specific syndromes are more common than in mID. Many of these syndromes include medical problems for which there is no treatment. However, the mortality rate in older age groups in which persons with certain syndromes (e.g., Down's) or life-shortening comorbidities have already died, approaches that of the general population (Janicki et al. 1999). In the only population sample study found (Patja et al. 2000), during a 35-year follow-up period people with mild ID shared a similar life expectancy to that in the general population.

Because of improving health and social welfare, the life expectancy of persons with intellectual disability is increasing along with that of the general population (Patja et al. 2000, Bittles et al., 2002, Perkins and Moran, 2010, Coppus 2013). In their study covering the time span of 1930–1980, Carter and Jancar (1983) noticed a marked increase in longevity among intellectually disabled persons living in hospitals. In persons with Down's syndrome the increase was 40 years, and in others over 30 years. There was a marked change in the causes of death during the study period.

Partly the change paralleled that in the general population, and partly the roles of specific causes of death (e.g. status epilepticus) diminished. The change was due to the introduction of new drug therapy, and better diet, care and environment for intellectually disabled people.

The prevalence of ID at an older age (65+ years) has been less than 0.3%. The general difficulty of including persons with mID in the study population may be exaggerated in old age. For a person of 65+ years, diagnostic assessments should have been performed decades previously – if not, it may be impossible to reconstruct the developmental progress afterwards in a cross-sectional study. A very long prospective study would be the best setting (Patja et al. 2000).

Table 6. Epidemiological studies of elderly persons with ID, in alphabetical order by author. Population sample study is shaded grey. SMR = standardized mortality rate.

Study, Country	Sample, Design, N	Outcome variables	Results	Remarks
Studies investigating mortality among persons with ID compared to general population				
Arvio et al. 2016, Finland	Register-based (deaths among persons receiving benefits from the SII with diagnosis of ID, compared with general population, time span 1996–2011). Total person-years in follow-up 378,987.	SMR	At age ≥ 60 years, SMR was 1.99 for persons with mID, and 2.07 for persons with sID compared with general population.	Only ICD codes F70–79 as inclusion diagnoses. Proportion of mID 40%.
Balakrishnan and Wolf 1976, Canada	Register-based (persons with sID in Canadian institutions 1966–1968, life expectancy compared with general population). n=739 (≥ 60 y)	Life expectancy	Life expectancy at the beginning of interval (m/f) was 14.3/13.2 years at 60–64 y compared with 16.8/20.6 years in general population, decreasing to 4.6/4.5 years at 75–79 y, compared with 8.4/9.9 years in general population.	Only sID
Bittles et al. 2002, Australia	Register-based (Disability Services Commission of Western Australia, 1953–2000, database used to calculate survival probabilities). N=8724	Median life expectancy	Median life expectancies 74.0, 67.6, and 58.6 years for people with mild, moderate, and severe levels of handicap.	Proportion of mID 55%
Carter and Jancar 1983, UK, England	Register-based (Stoke Park hospital group persons with ID 1930–1980). Total number of deaths 1383.	Mortality trends	Increased longevity, especially after 1960's. Average age at death during period (males/females) 1931–35: 14.9/22.0 1951–55: 29.2/36.3 1961–65: 34.6/46.7 1976–80: 58.3/59.8	No information on the levels of ID
Glover et al. 2016, UK, England	Register-based (Clinical Practice Research Datalink database for April 2010 to March 2014). 11.16 million person-years covered.	SMR, life expectancy	All-cause standardized mortality ratio for persons with ID was 2.8 at 65–74 y, 2.2 at 75–84 y, and 1.6 at 85–99 y. Their life expectancy at birth was 19.7 years lower than for people without ID.	GPs in England currently identify only around 0.5% of the population as having ID, suggesting that individuals with mild, non-syndromic ID are largely missed.

Table 6. *Epidemiological studies of elderly persons with ID, in alphabetical order by the author. Continued ...*

Study, Country	Sample, Design, N	Outcome variables	Results	Remarks
Janicki et al. 1999, USA	Register-based (death register, deaths of adults with intellectual disability 40 years of age and older who died during the period 1984–1993). Number of deaths 2752.	Average age at death	Average age at death for the persons with ID 63.3 / 66.9 years (m/f) compared with general population 65.8 / 74.1 years (m/f). During the 10-year study period, there was an average increase in mean age at death of 0.1% per year for institutional residents and 6% for adults living with their families, and a mean age decrease of 2% for adults living in supervised settings.	The results are consistent with other findings, that the life expectancy of successive generations of adults with ID, not otherwise compromised, will soon approach that of the general population.
Landes 2017, USA	Household-survey (Data from the 1986–2011 National Health Interview Survey–Linked Mortality Files). N=4812, with 69,460 person-years.	Relative mortality risk in different age groups	Relative mortality risk decreased from 2.53 at 60–69 y to 1.57 at 70–79 y, and to non-significant at 80–89 y	Individuals with a proxy or self-reported limitation in adaptive behaviour caused by an intellectual disability in the NHIS were designated as having intellectual disability. Diagnostic assessment was not described.
Lauer and McCallion, 2015, USA	Register-based (Data from US state intellectual and developmental disabilities service system administrative data sets and de-identified New York state Medicaid claims, 2009–2011). N=127,576 and 146,317 on average annually.	Average age at death	Average age at death for people in state intellectual and developmental disabilities systems was 50.4–58.7 years and 61.2–63.0 years in Medicaid data	The study population represented approximately 0.5% of the each state's population. The New York data represented approximately 0.7% of the state's population. No information on levels of ID.
McCarron et al. 2015, Ireland	Register-based (the National Intellectual Disability Database and the Census in Ireland in 2012). N=31,943.	Average age at death	The average age at death for males with intellectual disability was 17 years lower than males in the general population and 21 years for females respectively. For persons with mild/moderate ID the difference was 12–14 years, and for severe/profound ID 20–29 years.	Proportion of mID 30%
Ouellette-Kuntz et al. 2015, Canada	Register-based (a retrospective cohort study of mortality among individuals with intellectual and developmental disabilities in a region of Ontario). N=6054, with 29046 person-years.	SMR	SMR 1.7 / 2.1 (m/f) for 60+ y	Also included autistic spectrum disorders.
Patja et al. 2000, Finland	Population sample (representative stratified random sample of the whole of Finland, "Finland-in-Miniature" study, follow-up 35 y). N=2366, with 61,689 person-years.	SMR	People with mild ID share a similar life expectancy with that in the general population.	Proportion of mID 53%.

Table 6. *Epidemiological studies of elderly persons with ID, in alphabetical order by the author. Continued ...*

Study, Country	Sample, Design, N	Outcome variables	Results	Remarks
Tyrer et al. 2007, UK, England	Register-based (persons with moderate to profound ID living in Leicestershire and Rutland, between 1993 and 2005). n=2346	SMR	SMR 2.5 / 5.0 (m/f) for 60–69 y SMR 1.4 / 1.6 (m/f) for 70+ y Prevalence of sID 0.35% for 60+ y	Only sID
Studies on prevalence of ID in old age				
McConkey et al. 2006, Ireland and Northern Ireland	Register-based (the National Intellectual Disability Database/Ireland, local and national authorities for Northern Ireland). N=885/487 (Ireland/Northern Ireland) for 65+ years.	Prevalence	Prevalence 65+ 0.21% / 0.20 (m/f) Ireland, 0.26% / 0.19% (m/f) Northern Ireland	Proportion of mID 32% Ireland, 27% Northern Ireland
Ng et al. 2015, Sweden	Register-based (national register based on the Swedish Act concerning Support and Service for Persons with Certain Functional Impairments and the national death register, persons 55+ y, 2004–2012).	Prevalence	Prevalence of ID diminished steadily from 0.44% at 55–59 y to 0.065% at 80+ y. In all age groups the prevalence was higher among males, 23.4% at 55–59 y, and 20.2% at 80+ y.	The study population included individuals with ID, autism or pervasive development disorders. Diagnostic information not given.
Yen et al. 2013, Taiwan	Register-based (ID population structure compared with the general population). N=2800, 65+ years	Prevalence	Prevalence 65+ 0.13%. Distribution by age shown. (See Figure 6)	No information on the level of ID.

2.3.2 Low- and middle-income (LAMI) countries – children

Studies in low- and middle-income countries suffer from poorer quality than respective studies in high-income countries (Maulik and Darmstadt 2007, Jeevanandam 2009, Mercadante et al. 2009, Njenga 2009). Most are based on random household surveys with fairly small samples compared with studies performed in high-income countries. The tests used to screen samples are simple. Thus, validity may be questioned.

Seventeen studies containing information from 11 different countries are presented in this review (*Table 7, Figure 9*).

The prevalence of sID ($IQ \leq 50$) was generally higher and more variable in LAMI countries than in high-income countries (*Figure 8*). The median in this set was 0.66% (mean 1.00%, SD 0.67%, range 0.42–2.78%). These figures were more than twofold greater than in high-income countries, in several studies even threefold greater.

The prevalence of mID also showed great variation. The median in this set was 1.87% (mean 3.28%, SD 3.74%, range 0.4–14.0%). Also, these figures were almost threefold greater than in high-income countries. The proportion of mID of the total prevalence was very variable. The median in this set was 69% (range 31–90%).

Naturally, the total prevalence was very variable as well. The median in this set was 3.55% (mean 4.41%, SD 3.74%, range 0.90–15.6%).

Because prevalence figures for different ages were collated and the studies covered a limited age span, the association between prevalence and age could not be estimated.

Prevalence estimates were remarkably greater than in high-income countries. Besides methodological differences and problems, there can be true differences in prevalence due to poverty, a higher morbidity rate, poor diet and health care, lack of education etc. The intelligence tests may also give a too negative picture when they expect abstract thinking (Flynn 2007).

Table 7. Prevalence studies of ID in childhood and adolescence in low- and middle-income (LAMI) countries, in order of the year of publication. Population sample studies are shaded grey.

Study	Material	Methods	Results sID / mID / TotalID
Stein et al. 1987; Bangladesh,	Random household survey, ages 3–9 y, N=987	Ten-question interview, psychological examination using formal and informal techniques (no information on the tests), IQ ≤ 70 .	1.62% / 14.0% / 15.6% Proportion of mID 90%
Stein et al. 1987; Brazil,	Random household survey, ages 3–9 y, N=1058	As above	0.66% / 5.77% / 6.43% Proportion of mID 90%
Stein et al. 1987; India,	Random household survey, ages 3–9 y, N=1439	As above	2.78% / 1.25% / 4.03% Proportion of mID 31%
Stein et al. 1987; Malaysia,	Random household survey, ages 3–9 y, N=959	As above	1.25% / 0.94% / 2.19% Proportion of mID 43%
Stein et al. 1987; Pakistan,	Random household survey, ages 3–9 y, N=995	As above	1.51% / 2.11% / 3.62% Proportion of mID 58%
Stein et al. 1987; Philippines,	Random household survey, ages 3–9 y, N=1000	As above	0.50% / 0.40% / 0.90% Proportion of mID 44%
Stein et al. 1987; Sri Lanka,	Random household survey, ages 3–9 y, N=962	As above	0.52% / 0.73% / 1.25% Proportion of mID 58%
Stein et al. 1987; Zambia,	Random household survey, ages 3–9 y, N=1139	As above	0.44% / 2.63% / 3.07% Proportion of mID 86%
Islam et al. 1993; Bangladesh,	Random household survey, Urban slum/mixed urban-rural, ages 2–9 y, N=10,299 screened.	Ten-question interview, physician and psychologist (no details of the methods)	0.59% / 1.44% / 2.03% Proportion of mID 90%
Zuo et al. 1994, China	Population sample (all children born 1.7.1968–30.6.1982 and living permanently in Chang-qiao area), N=7682.	IQ was evaluated with standardized psychological tests.	Maximum rates for 10–14 y: 0.42% / 0.71% / 1.13% Proportion of mID 63%
Temtamy et al. 1994, Egypt,	Random household survey, ages 2–18, y N=3000	Screening:shortened Stanford–Binet test Ascertainment: Complete Stanford–Binet test.	0.83% / 1.87% / 2.70% Proportion of mID 69%
Durkin et al. 1998; Pakistan, Karachi	Population sample (random cluster household sampling 1988–1989, ages 6–9 y, 94% urban, 6% rural), N=6365.	Ten-question interview identified 14.7% suspects, psychometric testing (nonverbal scales from the 1985 revision of the Stanford–Binet IQ test), IQ ≤ 70 .	1.74% / 7.78% / 9.52% Proportion of mID 82%
Bashir et al. 2002; Pakistan, Lahore	Population sample (house-to-house survey to find all pregnancies, ages 6–10 y N=1607, in three poor, and one upper-middle-class area, birth cohort followed-up until 6–8 y of age), N=649.	Ten-question interview identified 132 suspects, psychometric testing (WISC-R and Griffiths).	Only mID: 6.2% Strong connection to socioeconomic background
Christianson et al. 2002; South Africa,	Random household survey (house-to-house basis by interviewing mothers or caregivers, ages 2–9 y), N=6692.	Ten-question interview, Griffiths Scale of Mental Development.	0.64% / 2.91% / 3.56% Proportion of mID 82%
Gustavson 2005; Pakistan, Lahore	Population sample (all pregnancies in geographically defined area, 6–10 y), N=1476	Follow-up every 3rd month up to 6 y, then twice a year by paediatricians and psychologists. Ascertainment methods not explained. IQ < 70 .	6.2% / 1.1% / 7.3% Proportion of mID 85%
Xie et al. 2008, China, six out of 31 provinces	Population sample (multi-phase, stratified, unequal proportional and cluster sampling), 0–6 y. N=60,124.	All screened for ID by the Denver Developmental Screening Test, positives tested by the Gesell Developmental Inventory.	0.42% / 0.61% / 0.93% Proportion of mID 55%
Karam et al. 2016; Brazil, southern city Pelotas	Population sample (live-born infants born in maternity hospitals in Pelotas in 2004. Follow-up at 3, 12, 24, and 48 months), N=4231.	At 48 months IQ test, Wechsler Preschool and Primary Scale of Intelligence (WPPSI)..	Total 4.5% / 4 y

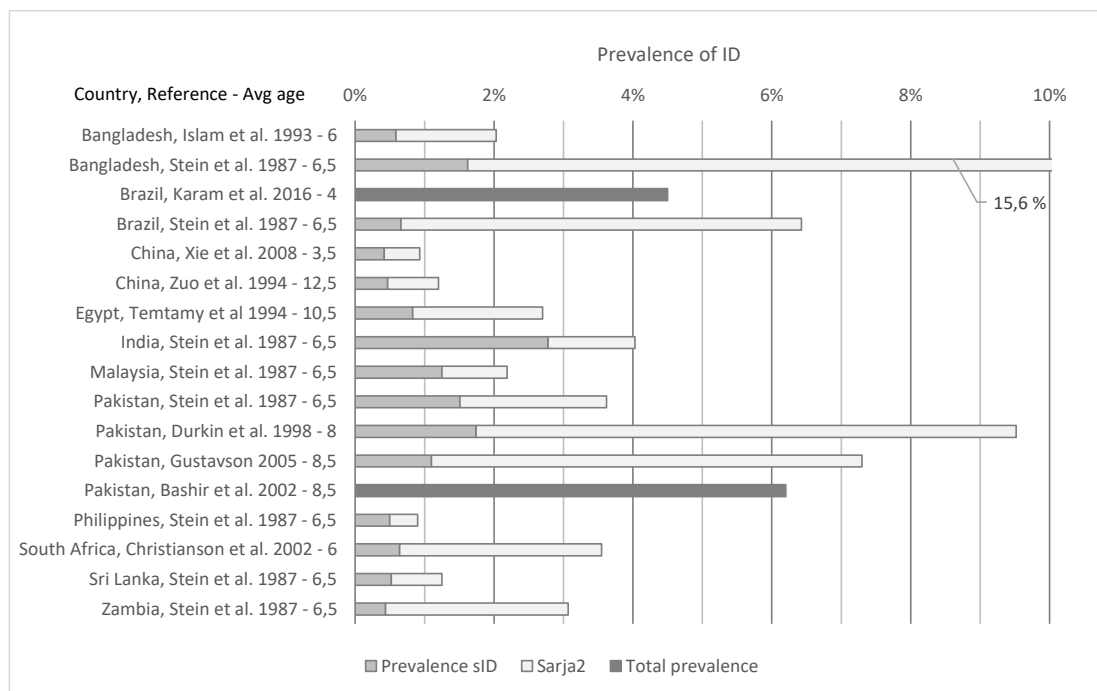


Figure 8. Prevalence of ID in childhood in low- and middle-income (LAMI) countries. Studies in alphabetical order by country.

2.3.3 Surveys and studies in Finland

For the purposes of comparison with the present study, I tried to trace all previously published available Finnish scientific studies and less rigorous surveys (*Table 8*). In addition, information on child disability allowance and disability pension was manually picked from SII statistical yearbooks (see also *Figure 9*).

During early decades of the 20th century, the study method was household surveys. Register-based studies became available when The National Pension Institute was founded in 1937 and started to allow disability pensions in 1942. To support development in legislation in the care for persons with ID, the population sample study "Finland-in-Miniature" was undertaken in 1962 (Amnell 1966, Ruoppila 1966, Tarvainen 1966). It was remarkable, one of the very first internationally, and still the only one with very long follow-up, 35 years (Ruoppila and Iivanainen 2011). Despite higher prevalence estimates in scientific studies, the administrative estimates have remained at a lower level.

During the 1980's service registers for persons with ID were kept in communes, and the data was summed up nationally. Due to lack of resources for reliable updating, the registers were discontinued. The data was used in a scientific study where the register prevalence rate in 1987 was compared with that of the Northern Finland birth cohort 1985/86 (Gissler et al. 2000). At seven years of age, both data collection methods gave equal results for the cumulative incidence of intellectual disabilities.

Table 8. Published surveys and studies on the prevalence of ID in Finland. MOD = Modified education for developmentally delayed, borderline and mild ID, TR1 = training education for pupils with ID, TR2 = training education for pupils with most severe ID.

Study	Year	Method	Results
Governmental Committee on Psychiatric Care [Mielisairaanhoitokomitea] (see Tarvainen 1966)	1906–1907	Household survey, no testing. Probably mostly sID.	Prevalence in rural communes 0.34%, in urban communes 0.13%, in total 0.31%
Governmental Office for Social Studies [Sosiaalinen tutkimustoimisto] (see Tarvainen 1966)	1935–36	Household survey, psychological testing of suspected cases, no information about the tests or diagnostic criteria available	sID in rural communes 0.48%, in urban communes 0.28%, in total 0.44%. Maximum prevalence among 10–14-year-olds 1.2%, and among 15–19-year-olds 0.54%.
Survey by professor Martti Kaila (see Tarvainen 1966)	1942	Statistics of invalidity pension by National Pension Institute concerning 22–58-year-olds. Diagnostic criteria not known	Average prevalence of sID across all ages 0.52%
The Office of District Physician for Lapland [Lapin lääninlääkärin toimisto] (Tarvainen 1966)	1958	In the district of Lapland, 1–15-year-olds screened by district nurses, and diagnosed by the district physician in ca. 73% of the cases. No information about diagnostic criteria.	Average prevalence of sID 0.3%
Governmental Office for Social studies [Sosiaalinen tutkimustoimisto], see Ruoppila 1966 in Table 3.	1962	"Finland-in-Miniature", 9.4% sample of the whole of Finland, 2–64-year-olds	Total ID 0.66%, IQ 50–69: 0.30% IQ 20–49: 0.24% IQ 0–19: 0.10%
Birth cohort study in capital of Finland, see Amnell 1974 in Table 3.	1969	All children born in 1955, follow-up until 13.4 years	IQ 0–69: 0.92% IQ 0–75: 1.61% IQ 68–85: 2.52%
Cohort study in Eastern Finland, see Kääriäinen 1987 in Table 3.	1985	All 8-year-old children in the district of Kuopio	Total ID 1.38%
Northern Finland Cohort Study 1, See Rantakallio and von Wendt 1986 in Table 3.	1986	All children born in two northern districts in Finland in 1966	sID at 11.5 y 0.60% (95% CI 0.46–0.74) mID at 11.5 y 0.50% (95% CI 0.38–0.63)
Northern Finland Cohort Study 2, See Heikura et al. 2003 in Table 3.	2003	All children born in two northern districts in Finland in 1985/1986	sID at 11.5 y 0.38% (95% CI 0.25–0.50) mID at 11.5 y 0.75% (95% CI 0.57–0.92)
Statistics from Registers of Social Insurance Institute (Annual Yearbooks by SII 1972–2015)	1972–2015	Child Disability Allowance for 0–15-year-olds, ID as the main diagnosis (ICD-10: F70–F79)	1972: 0.15% 1998: 0.30% 2015: 0.30%
Statistics from Registers of Social Insurance Institute (Annual Yearbooks by SII 1961–2000)	1961–2000	Invalidity Pension for 16–64-year-olds, ID as the main diagnosis (ICD-10: F70–F79)	1961: 0.47% 1972: 0.60% 2000: 0.68%
The National Board of Social Care: Task force for the development of special care for ID (Sosiaalihuoltus 1986)	1986	Administrative estimate, average for all ages	Prevalence of ID 0.64%
Ministry of Social and Health Affairs: Task force for the development of service systems (Matikka 1992)	1992	Administrative estimate, average for all ages, based on statistics of disability pension and special education.	Prevalence of ID 0.54%
Statistics of special education in 1994–95. (Virtanen and Ratilainen 1996). All pupils N=587,800	1994–95	Modified education for developmentally delayed, borderline and mild ID (MOD); training education for pupils with ID (TR1); training education for pupils with most severe ID (TR2)	Prevalence in MOD 1.70%, n=10,071 Prevalence in TR1 0.50%, n=3087 Prevalence in TR2 0.12%, n=718
Statistics of special education in 1998 (Statistics Finland 1998). All pupils N=581,900	1998	As above	Prevalence in MOD 1.70%, n=9873 Prevalence in TR1 0.55%, n=3229 Prevalence in TR2 0.15%, n=866
Ministry of Social Affairs and Health. Report on the Services for people with intellectual disabilities in 2004 (Kumpulainen 2007)	2007	Administrative estimate, average for all ages. Questionnaire sent to all communes and special care districts	Total ID 0.67% Among services 0.52%

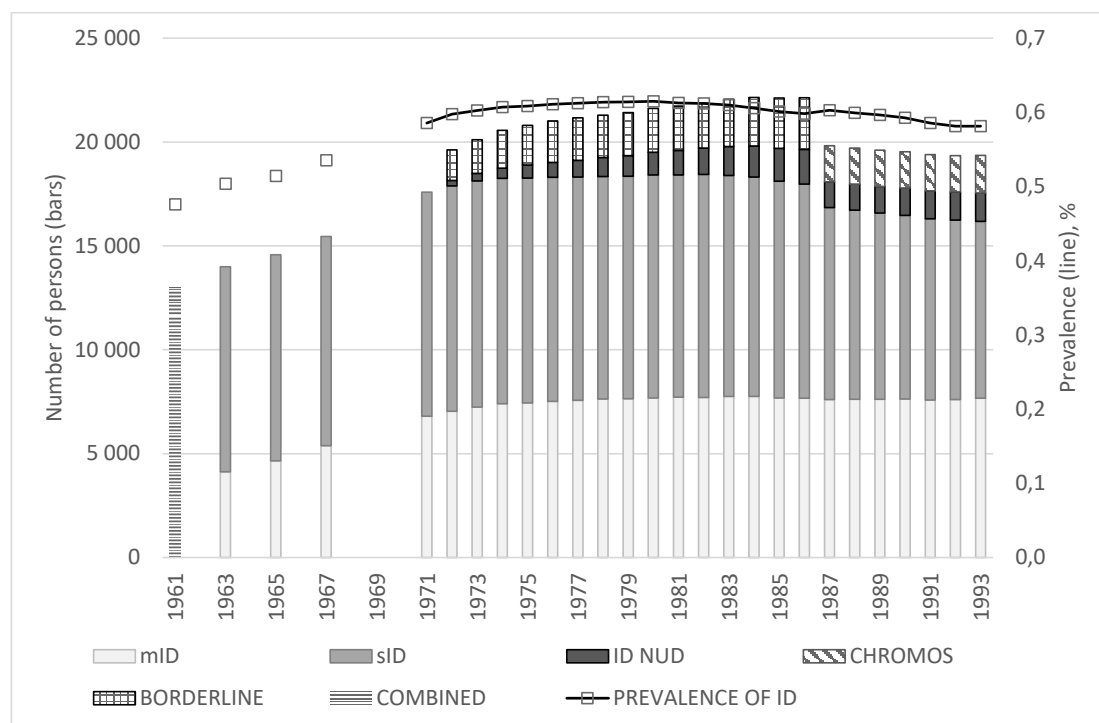


Figure 9. Register-based prevalence of ID among 16–65-year-olds, manually picked from SII statistical yearbooks. Disability pension based on borderline ID was available in 1972–1986, chromosomal disorders as aetiology became available with ICD-9 in 1987. Concerning the year 1961, information on levels of ID was not available. Prevalence of ID (line) does not include borderline ID.

2.3.4 Why is there such great variation in prevalence estimates?

Despite the long tradition of epidemiological research in ID, it is difficult to draw a comprehensive picture. The studies are very variable about materials and methods, which are often incompletely described. The definitions and criteria used vary. It is particularly difficult to verify how the criterion of adaptive behaviour has been considered in the assessment. A remarkably large proportion of the studies have been carried out without describing explicitly the socioeconomic structure of the study population, although its explanatory power in prevalence estimate differences has repeatedly been shown.

There is a risk of over-inclusiveness in ethnic and language minorities, especially when verbal abilities are emphasized in the assessment.

As previously noticed, the prevalence of sID is more consistent than that of mID. Severe ID is more often due to some recognized medical syndrome. As there are so many syndromes having an influence on the development of cognitive performance, it is somewhat surprising that the prevalence of sID is so consistent. In contrast, the prevalence of mID varies to a wide extent. It has been suggested that while sID is related to medical syndromes, mID is more connected to the socioeconomic situation.

2.3.4.1 *Role of study design, case-finding, diagnostic protocol and cut-off values*

In this review, in 41 studies concerning under 18-year-olds in high-income countries the total prevalence of ID was reported. In 23 studies the study design was population sample, in 18, register-based. The median prevalence of ID did not much differ between these groups, 1.12% (population sample) vs. 1.10%; neither did the median prevalence of sID, 0.39% vs. 0.42%. The median proportion of mID in the study populations was similar too; 55.0% vs. 56.0%.

In a systematic review, the data was analysed according to study design, comparing prevalence estimates in cross-sectional and cohort studies. The figures were 0.97% (95% CI 0.87–1.06) in cross-sectional studies, and 1.32% (95% CI 1.07–1.57) in cohort studies (Maulik et al. 2011). This analysis covered a mixture of studies from both high-income and LAMI countries.

Analysing ten Scandinavian prevalence studies for ID, Kebbon (1987) concluded that case-finding methods are the most decisive about prevalence figures in epidemiological studies, especially concerning mild mental retardation. While the prevalence of sID appears to be consistent, the total prevalence estimate seems to depend largely on the inclusion of persons with mID in the study group, as indicated in *Figure 4*.

One study has been specifically aimed at defining the role of taking adaptive behaviour into the diagnostic algorithm (Obi et al. 2011). The study evaluated the effect of incorporating adaptive functioning (AF) data on overall intellectual disability prevalence according to sociodemographic, economic, and severity characteristics. Among 1595 children aged 8 years who met the study's intellectual disability surveillance case definition of $IQ \leq 70$, prevalence estimates showed few substantive changes when incorporating AF data. The authors concluded that use of IQ data alone appears to be sufficient for measuring population intellectual disability prevalence. Still, one cannot exclude the possibility that if the prevalence of ID were on a higher level than here (1.2%), as it has been in many studies, over-inclusiveness might be present.

The role of IQ cut-off values has also been evaluated in a few studies. Simonoff et al. (2006) reported prevalence ranges in connection with an IQ of 70 ± 3 in a deprived, multicultural population in a single borough in England with a high prevalence of ID. The range for boys was 6.3–13.4% and for girls 8.0–13.6% (calculated by the current author from Figure 1 in Simonoff et al. 2006). In an ethnically diverse population in Arizona, USA, Reschly and Jipson (1976) reported a prevalence estimate of 3.5% using an IQ cut-off value of ≤ 69 , and 7.5% when using $IQ \leq 75$. Thus, IQ cut-off points may have a considerable influence on prevalence estimates. Most prevalence studies have concerned $IQ < 70$ or ≤ 70 .

2.3.4.2 *Relationship with socioeconomic and ethnic factors*

The association between the prevalence of ID vs. socioeconomic status and race of the family was studied in the Prospective Perinatal Project in the USA (Broman et al. 1987). The association was significant (*Figure 10*). Total prevalence figures were 1.68% for whites and 5.27% for blacks at seven years of age. In white and black populations, the prevalence of sID was at the same level. However, in the lowest socioeconomic group it was twice as high as in other SES groups. In contrast, the prevalence of mID was at a higher level in the black population, and it varied with socioeconomic group more strongly. The study had a prospective longitudinal design and a large sample size (36,851 children). The authors pointed out that the mothers in this project received excellent medical care, while in the general

population of lower-class mothers such care is often lacking. Therefore, the differences between the groups might have been even more extreme without good medical care.

In a similar population sample study (Camp et al. 1998) the findings were replicated. Total prevalence figures were 1.7% for whites and 5.5% for blacks at seven years of age. Socioeconomic status of the family accounted for 44–50% of intellectual disability and a low level of maternal education accounted for 20%. Similarly, in Western Australia, a marked difference in the prevalence of ID was noted between Caucasian and aboriginal populations in two studies (Leonard et al. 2003, Bourke et al. 2016) (see page 39).

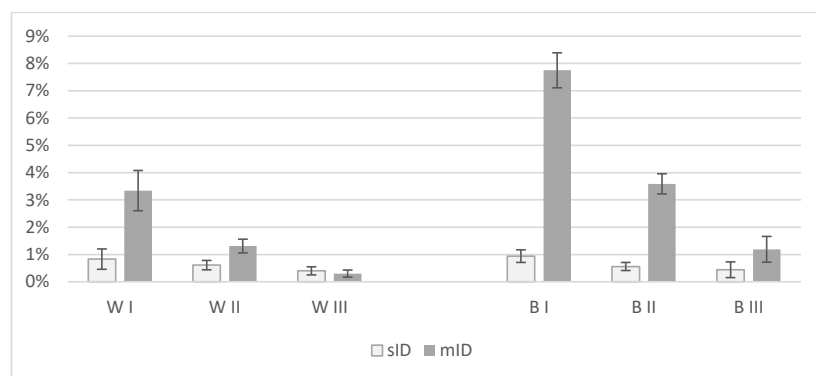


Figure 10. Association between prevalence of ID (sID and mID), race and socioeconomic status. W = white, B = black, I–III refer to socioeconomic group, I being the lowest. Whiskers are 95% confidence intervals calculated by the author (H.W.) from the original data (Broman et al. 1987).

A clinical and epidemiological population sample study of mental subnormality was carried out in a geographically defined area in Scotland (Birch et al. 1970). The relationship between the prevalence of ID and social class was analysed (Figure 11). In this study, too, only the prevalence of mID was consistently connected to socioeconomic status.

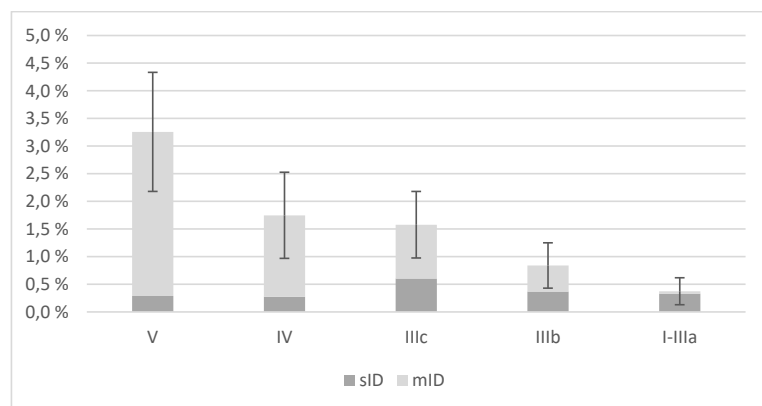


Figure 11. Prevalence of ID in the administratively classified group according to severity of ID (sID, mID) and socioeconomic status. In this data mID = IQ 50–75. Whiskers are 95% confidence intervals for the summed (sID

+ mID) values calculated by the author (H.W.) from the original data (Birch et al. 1970). Social classes are I–IIIa = Non-manual, IIIb = Journeyman and artisan, IIIc = Other skilled manual, IV = Semiskilled manual, V = Unskilled manual.

In Northern Finland, a one-year birth cohort was studied twice, 20 years apart, in 1966 and 1985/86 (Heikura et al. 2008). The risk populations were 12,058 and 9,432 respectively. Despite an interval of 20 years between the cohorts, the main indicators of socioeconomic disadvantage and maternal multiparity remained as having the largest impact on the incidence of intellectual disability, especially mID. In another register-based study covering all of Finland, the mother's occupation at the time the child was seven years old was highly correlated with the prevalence of ID (Gissler et al. 1998). The findings were not explained by perinatal health.

Associations between socioeconomic status, IQ and aetiology of ID were studied in Akershus county, in which economic status is ranked as the second highest in Norway (Strømme and Magnus 2000). Even here, lower socioeconomic status increased the risk of mID. Drews et al. (1995) noted that the relationship between socioeconomic background and prevalence of ID was more pronounced for isolated ID, i.e. without other neurological conditions.

The role of socioeconomic factors has also been studied in LAMI countries. In a study in Karachi, Pakistan, lack of maternal education (none vs. some) was the strongest of the 13 studied background factors associated with the prevalence of both sID (2.7% vs. 0.8%) and mID (9.1% vs. 3.0%) (Durkin et al. 1998).

In a review (Bergen 2008) ID was named as one of the most common causes of disability and one of the most prevalent neurologic disorders globally. Several factors connected to poverty may contribute to neurodevelopmental disabilities, such as protein/energy malnutrition, dietary micronutrient deficiencies, environmental toxins, and lack of early sensory stimulation or the ability to profit from it. Tropical diseases such as parasite infestation with resultant anaemia, malaria, and other infections are major contributory causes. It was concluded that reduction of poverty and its effects would reduce the present and future burden of intellectual disability and cognitive dysfunction, especially in LAMI countries. On the other hand, families supporting a child with intellectual disabilities and adults with intellectual disabilities are at increased risk of experiencing poverty due to the financial and social impact of caring, and exclusion of people with intellectual disabilities from the workforce (Emerson 2007).

Prevention of intellectual and other disabilities is in many ways tied to the plentitude of functions of a well-organized society (GBD 2017). It is easiest to understand the role and prerequisites of general hygiene and specific prevention programmes such as vaccination. However, many other societal domains also are important. Safety in traffic, prevention of drowning accidents, prevention of fires, good care of poisonous materials – all these functions together have immense influence. When they have become integrated in society, their importance can be forgotten, but the differences become obvious in comparison between high- and low-income countries, or subpopulations. Where general security is low, with or without open conflicts or wars, the risk of abuse of children is high, with resulting traumatization, deprivation and even pseudoretardation.

2.3.4.3 Exceptionally high prevalence figures and the role of reference values

The theoretical prevalence of persons with an IQ of more than 2 SD below average is 1.94% according to a normal distribution pattern (2.28% for the other commonly used cut-off point of ≥ 2 SD below the

mean). Keeping this fact in mind, it is astonishing that the great majority of studies report much lower prevalence figures. In this literature review there were five studies from high-income countries where the total prevalence in childhood was close to or above 1.94%: in UK/Scotland 2.74% (Birch et al. 1970), UK/England 2.52% (Rutter et al. 1970), in South Carolina, USA 4.16% (McDermott 1994), in Taiwan 2.80% (Hou et al. 1998), and in UK/England 5.8–10.6% (Simonoff et al. 2006, not in *Figure 2*). In France, the prevalence of mID was 1.80%, and when added to the median prevalence of sID (0.42%) it also reaches >1.94% (David et al. 2014).

Are these to be considered exceptional, or the few ones with reasonable and reliable results?

In the study carried out by Birch et al. (1970) the cut-off used was $IQ < 75$, which partly explains the high figure. In McDermott's study (1994) the definition of ID was based on the South Carolina State Department of Education Handbook, written for educational purposes. It is uncertain how well the definition matches those of AAMR/AAIDD. These two studies were among the three in which the proportion of mID of all ID was above 80%. In the paper by Hou et al. (1998) no details were given of the "nationwide screening program", nor were there details of the diagnostic assessment, but the IQ cut-off was ≤ 70 .

In the study by Simonoff et al. (2006) there were several uncertainties, such as a vague definition of the population, and scoring of the group and individual tests. The population was ethnically more diverse than average in the UK. The classification of ID relied only on the IQ score, not on adaptive behaviour.

The study reported by David et al. (2014) was a population sample study, with thorough clinical judgement. The methods are described in detail, and it is difficult to see clear biases.

The Isle of Wight Study (Rutter et al. 1970) is exceptional in one respect: the tests of IQ were normed in the study population. In this case, the logical result is to find the theoretical proportion according to the normal distribution, 1.94%. Rutter et al. found a prevalence value of 2.53% (95% CI 1.89–3.16%). The proportion of mID of all ID was above 80%. If the IQ had been calculated based on the WISC manual at that time, the prevalence rate would have been 1.46% (95% CI 0.97–1.94%, calculated by the current author) (Rutter et al. 1970, page 40). Rutter et al. discuss these findings, concluding that the child population on the Isle of Wight may be more intelligent than the reference population, normed based on the WISC test at the time.

The extensive study by Rutter et al. (1970) raises the serious question of whether or when the tests should be standardized in the study population. Only one other study could be found where this was done. Kääriäinen (1987) screened whole birth cohorts at the age of 8–9 years in the district of Kuopio, Finland; those born in 1969–1970 and 1971–1972. The tests were normed by using a random sample of 101 subjects. The prevalence of ID ($IQ \leq 70$) was 1.38% (95% CI, 1.18–1.58%, calculated by the current author). The prevalence fell much short of the theoretical value, 2.25%, which was not discussed by the author, and may mostly be a result of too small a population in the standardization process. In contrast to the study by Rutter et al., this study did not involve published norms for IQ tests in comparison.

2.3.4.4 *Development of general health care, specific aetiological factors, and preventive measures*

There are numerous factors which have changed the epidemiology of ID throughout the decades and may bring out differences in prevalence between different geographic areas or segments of any society.

Many infectious diseases that have ID as a possible sequela (e.g. human cytomegalovirus, rubella, toxoplasmosis, herpes, HIV-1) have been controlled, either via prevention (better general health, better hygiene, vaccination) or treatment (e.g. congenital syphilis) (Obladen 2013). Nevertheless, large differences exist between populations, and new infectious epidemics may appear, such as the zika virus in 2015 (McKenzie et al. 2016).

Better maternal care during pregnancy and delivery has been important in high-income countries, but effort is still needed in less high-income countries. Rh-immunization and congenital hypothyreosis are now well-known risks that can be prevented or treated in a timely manner. Exposure to toxic chemicals, either acute disaster or chronic contamination, presents a continuous threat in any society.

Intrauterine exposure to ethyl alcohol seems to be a serious and increasing problem. May et al. (2018) performed a study on the prevalence of alcohol-spectrum disorders among first-graders in four US communities. Using active-case ascertainment methods in a cross-sectional design between 2010 and 2016, the estimated prevalence ranged from 1.1% to 5.0% using a conservative approach. In Australia, maternal alcohol use disorder is the leading known risk factor of intellectual disability, with no identified genetic origin (O'Leary et al. 2013).

2.3.4.5 *Consanguinity*

In many countries with small isolated populations, often with a founder effect, some rare syndromes become apparent, as in northern Finland (de la Chapelle 1993). In Denmark higher prevalence rates have been noticed on smaller islands, where consanguinity is more common (Dupont 1989). In Pakistan, the common tradition of marriages between cousins has the same influence, even in a large non-isolated population. The elevated level of endogamy has led to an increased prevalence of genetic disorders, including autosomal recessive intellectual disability (ARID), with an average of 1.1 cases of severe ID and 6.2 cases per 100 live-births of mild ID (Grozeva et al. 2017).

2.3.5 **Changes in prevalence with age**

The results of several longitudinal studies have suggested that levels of intelligence are essentially stable throughout the life span (Mackintosh 2011) – people's IQ scores after the age of 10 remain relatively (although far from perfectly) stable for much of the rest of their life.

Hagberg et al. (1987) studied changes between two points of time in prevalence rates and distribution of IQ values in intellectually disabled children resident in Gothenburg, Sweden. The same group of almost 2500 pupils was examined both in 1978 (8–12-year-olds) and in 1984 (14–18-year-olds). The changes that occurred in IQ were mainly downwards. Thirteen of the 91 in the mID group in 1978 had moved to sID, three climbed to borderline, and 19 newcomers from borderline were identified as mID.

According to the definition, ID manifests itself before age 18. In some studies, higher prevalence figures have been obtained later, and in others vice versa, where a marked drop has been observed when reaching adulthood. If IQ remains relatively stable with age, then the changes would be due either to changes in adaptation or in attrition.

Granat and Granat (1973) discussed why in several previous studies the prevalence of ID seemed to drop between 14 years of age and adulthood. One possible reason was lack of proper test norms for adults, whereas they were very accurate for school populations. Another explanation was related to differences in accessibility between those populations. Their study targeted a representative sample of 2000 19-year-

old males registering for military service. After a brief intelligence test (for the whole age cohort), 217 men with the lowest scores in this sample were given a more comprehensive intelligence test, which was standardized in Sweden. No one in this group had previously had a diagnosis of ID. The test group was divided into two subgroups based on social adjustment according to defined criteria. The same test procedure was carried out in a comparison group of persons selected from two special hospitals for ID persons with special antisocial difficulties in adjustment. All three groups had similar average results in the psychological tests of IQ. The estimate for the prevalence of ID in the 19-year-olds registered for military service was 1.50%. Together with the 0.71% who were not enlisted because they were already diagnosed as having ID, the prevalence of 19-year-old men in Sweden fulfilling the psychometric criterion for ID was estimated to be 2.21%. Thus, a sizable number of men in this age cohort had gone through school without being labelled as intellectually disabled, but half of them were noted to have failed in social adaptation after school, and would be needing support sooner or later (Granat and Granat 1973).

Likewise, in the Netherlands, Stein et al. (1976) carried out a population sample study of 19-year-old men born in 1944–1947, using data from military records. Three inclusion criteria were used: special education for persons with ID, IQ tests, and previous ICD-9 diagnosis of ID. In sum, 3.0%–6.1% were evaluated as having ID.

Richardson et al. (1984) followed up children born during a five-year period until age 22, resident in a British city, and administratively defined as intellectually disabled. Age-specific prevalence rates ranged from 0.25% at age five to 1.5% at age 11. Almost three-quarters of those who had been at a school for educable, intellectually disabled children received no services in the young adult period.

2.3.6 Trends in time

While there have been epidemiological studies of moderate quality for decades, is there any trend in the incidence/prevalence of ID over time?

Registers for special services might be a good source of material to investigate time trends. The procedures used should be very stable – find eligible persons and accept them into the register – so that the only factor to bring out changes in the administrative prevalence would be real changes in the incidence of ID.

In Ireland, The National Intellectual Disability Database (NIDD) presents such a possibility. The database was founded in 1995. It is intended to provide a comprehensive and accurate information base for decision-making in relation to the planning, funding, and management of services for people with an intellectual disability. The objective is to obtain this information for every individual known to have intellectual disability and assessed as being in receipt of, or in need of, an intellectual disability service. In practice this means that only persons with sID are registered. The NIDD Committee publishes annual statistical reports. In the 2016 report (Doyle and Carew 2016), prevalence figures were compared with those in 1996, and with figures at two other time points (1975 and 1981) from another national source. As regards total prevalence there was no change from 1975 to 2016 (0.38%–0.36%–0.42%–0.36%). However, the prevalence had decreased in younger age groups (e.g. 0–14 years: 0.40%–0.32%–0.29%–0.29%) and increased in older ones (e.g. 54+ years: 1.71%–1.51%–2.11%–2.7%). It seems that the incidence has decreased, but the "wave of prevalence" travels through all age groups.

Richardson (1989) suggested in 1989 that there had been a decreasing trend in the incidence of ID during the prior three decades, based on analysis of studies from the US and Europe. During this study period both epidemiological interest started growing and service systems for persons with ID began to develop, as regards the most severely handicapped persons. In connection with these important changes one should be cautious in identifying possible changes in the incidence of ID. The lack of a clear definition of intellectual disability in many studies and the lack of a description of the methods used in selecting subjects make it difficult to determine to whom the results apply.

In Northern Finland two cohort studies were carried out, with a 20-years time difference (1966 vs. 1985/86), using the same methodology. There was no change in the total prevalence of ID at the age of 11.5 years (1.10% vs. 1.12%). There were, however, changes in developmental levels. In the former cohort the ratio of sID:mID was 70:59, whereas in the latter it was the opposite, 35:70 (Heikura et al. 2003). The authors discuss that there might be a multitude of explanations for the variation, such as inconsistent criteria for case definition, varying age of cohorts, ascertainment methods and definitions. However, because of the representative population samples and data collection from the same geographical area, using the same data sources, and the same criteria for case definition, they are inclined to believe that these changes reflect true changes in prevalence.

There may be several parallel trends. One is a decreasing trend in the prevalence of ID with socioeconomic development, which concerns some countries (and others for some segments of the population), e.g. advances in medical care, especially pre- and postnatal care, allocation of educational services for children with intellectual disability, and improved service systems in society in general (Heikura et al. 2003). In contrast, in less developed populations, especially with malnourishment following famines and disease epidemics, developmental disabilities will occur at a higher level. Even in high-income countries there is a continuous risk among marginalized people of increasing incidence of ID due to lack of preventive health care, lack of good nutrition and increasing alcohol consumption, among other risks. There is very little research on such long-term changes but the big picture may be gathered from many sources.

2.3.7 Borderline intelligence and ability to cope

In the above, *reliability* of the measurement of IQ has been mainly discussed, but the *validity* of cut-off values (in IQ and adaptive behaviour) is another aspect. Those with IQ scores slightly below do not much differ from those whose scores are slightly above, although with a larger IQ difference the difference in everyday performance is marked. Again, recently some authors have raised the concern that persons with an IQ in the borderline region (70–85) may become marginalized (Hassiotis 2015). Their adaptive abilities may not compensate for the lower-than-average intellectual performance (Peltopuro et al. 2014). According to the normal distribution pattern, as much as 13.2% of the population falls into this category.

In DSM-IV-TR, borderline intellectual functioning (BIF) is defined by IQ in the 71–84 range. In DSM-5, IQ boundaries are no longer part of the classification, leaving the concept without a clear definition, which according to Wieland and Zitman (2016) is one of the least highlighted changes in DSM-5. The diagnosis of BIF is not included in the current version ICD-10 either. In the 1975 revision (ICD-9) it was included with the code V628A.

In Finland, borderline intelligence was accepted as a diagnosis for invalidity pension (if other criteria were fulfilled, as with other relevant diagnoses) according to ICD-8 and ICD-9, but the diagnosis was cancelled in the next version, ICD-10, which came into use in 1996. According to statistics of the National Pension Institute, in 1972–1986, 1467–2490 persons received invalidity pension after this diagnosis (see *Figure 9*). These figures were 8.1–12.7% of the numbers receiving invalidity pension after a diagnosis of ID, showing an increasing time trend.

Wieland and Zitman (2016) have pointed out that BIF is an important and frequently unrecognized comorbid condition relevant to the diagnosis of all psychiatric disorders. They describe that in the Netherlands, individuals with BIF and comorbid psychiatric disorders are eligible to the same specialised mental healthcare services as people with ID, while it is doubtful whether general mental healthcare services can deliver the same adequate care.

Whitaker (2013) pointed out that there is only a loose relationship between being eligible for a diagnosis and being able to cope. Besides measurement errors in assessment, there may be several other important factors behind misclassification. The most important may be comorbidity. A person may have borderline IQ (not being eligible for a diagnosis of ID), but due to autistic spectrum disorder, mental illness, or sensory problems his/her ability to cope may be low. One may reason that the correct implication in policy would be not to build services which too narrowly focus on one problem. Another problem concerns those normative demands that appear later in life after the developmental period up to 18 years of age. One may have been able to cope and score on adaptive behaviour measures well enough during childhood and adolescence, but the person may fail later in life in critical intellectual tasks such as providing adequate child-care or protecting oneself from exploitation, and would need services specialised for ID persons. Modern society can make new demands on its inhabitants, and many such demands require abstract thinking that can be challenging for a person with ID (Whitaker 2013).

In the USA in 1969, the President's Committee on Mental Retardation² (PCMR) co-sponsored a conference under the title "the six-hour retarded child", meaning that a proportion of those labelled as having borderline or mild ID in academic settings fared reasonably well, if not on par with their peers, outside the academic setting (President's Committee on Mental Retardation 1969). The focus was on concerns over cultural bias and the arbitrariness of labels in the assessment of children with mild intellectual impairments.

Thirty years later, the PCMR revisited the cohort in its report entitled the "Forgotten Generation" (President's Committee on Mental Retardation 1999). There was a suspicion that those who were not labelled as mentally retarded, and therefore not receiving services, but who had marked cognitive limitations, did not cope so well as expected. Fujiura (2003) analysed data from the 1994 and 1995 National Health Interviews on Disability, which was the first national-level household survey specifically targeting persons with disabilities across all ages. In this retrospective study, three groups were compared with the general US population: those with 1) a specific learning disability, 2) mild intellectual disability, and 3) mental retardation (in the meaning of the PCMR). The proportions of respondents in each group with an unmet need as regards clinical and support services were calculated. These proportions were nearly identical in the labelled and unlabelled groups with cognitive impairment

² The term used was 'mental retardation' to refer to persons with ID who have been accepted to receive services ('labeled'), whereas mild ID referred to persons with cognitive impairment without being included in services for ID ('non-labeled').

(46.8% vs. 46.4%), and lower among persons with specific learning disabilities (34.3%). Two groups, mild intellectual disability and mental retardation, were very similar to each other in different support domains: communication, self-care, home living, social skills, community use, and work. The findings support the hypothesis of a "forgotten generation". Because of the low response rate, the results must be considered tentative.

In a project starting in 1977 in Finland and lasting for more than two decades, with increased efforts, persons with disability and incapacity for work were thoroughly assessed (Taipale 2001). This included detailed anamnestic information from early development to present performance, from school certificates, health records, reports from social care, employment offices, previous employers, the army, and often also custody care, and prison, and investigations to map unrecognized health problems and possible neuropsychological problems. Around a thousand individuals were assessed as being eligible for invalidity pension, and among the others the most common diagnoses were borderline intelligence, other neuropsychiatric conditions – at that time 'minimal brain dysfunction' (MBD) nowadays mostly ADHD, and severe dyslexia. According to the experience from the project, about 10–15% of aging persons with incapacity to work would be eligible for invalidity pension, which means 10,000–15,000 individuals. This is sizable number compared with estimates of the prevalence of ID (see *Table 8*).

Greenspan (2017) has expressed a contrary opinion on the relevance of BIF. He points out that full-scale intelligence quotient (IQ) is an outmoded concept even as regards ID – DSM-5 states that measures of 'executive functioning' (reasoning, planning, consequential thinking, attention, self-regulation, and so on) are often more meaningful than full-scale IQ as diagnostic indicators of ID. The coping abilities of those with borderline intellectual functioning might thus be better described as being related to cognitive problems of various subtypes.

2.4 SUMMARY OF THE LITERATURE

Despite a long tradition of epidemiological research into ID, it is difficult to draw a comprehensive picture of its prevalence. The estimate depends heavily on study design, characteristics of population, screening and assessment methods, criteria, and cut-off values of the tests used. Most of the studies are register-based, where information concerning psychological tests and their versions either has not been available or has been too cumbersome to find. The same holds true concerning assessment of adaptive behaviour. In only a few studies has there been a mention about which assessment methods were used for adaptive behaviour (Obi et al. 2011). Concerning study populations, the great majority of publications have not described the socioeconomic status of the study population, although it has a crucial influence on the prevalence figures. Likewise, the proportion of cases of mID has a major influence on the estimate of total ID prevalence. It seems that in previous meta-analyses not enough emphasis has been given to this fact (Roeleveld et al. 1997, Maulik et al. 2011).

Prevalence estimates of sID in high-income countries seem to have been fairly stable at around 0.3–0.4%. In contrast, prevalence estimates of mID are very variable, and largely connected to socioeconomic status. In LAMI countries the epidemiological study methods have not been as rigorous as in high-income countries, household survey being the most frequently used method. The prevalence estimates are very variable, even for sID. Population sample studies give higher estimates.

There is a risk of over-inclusiveness in ethnic and language minorities, especially if verbal abilities are emphasized in the assessment. In contrast, if adaptive behaviour and practical skills are emphasized at the expense of cognitive abilities, there is the risk of exclusion from necessary services for persons with ID, who may become marginalized at some later age with new normative demands.

3 AIMS OF THE STUDY

The purpose of this study, reflected in Articles I–IV was to estimate the prevalence of ID in Finland using existing health and social-care registers. Specific aims were as follows:

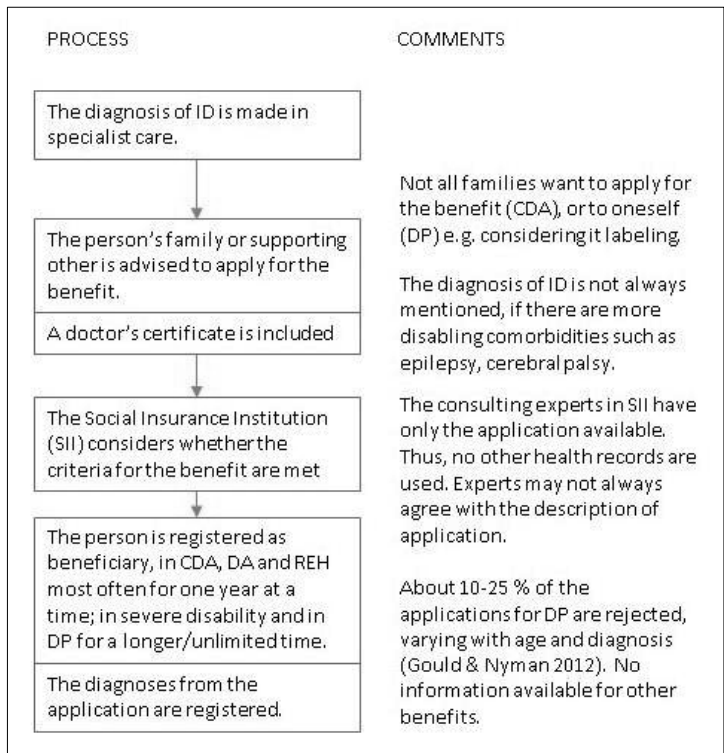
1. To estimate the prevalence of ID in the whole population in Finland, using multiple health and social-care registers (Article I)
2. To estimate the prevalence of ID in one-year age cohorts using multiple health and social-care registers (Article II)
3. To estimate the prevalence of ID in the elderly population using mathematical correction of underrepresentation in previous data (Article III)
4. To estimate the prevalence of ID using cumulative analysis throughout childhood and adolescence, with one health-care register (Article IV)

4 METHODS

4.1 STUDIES I–III

In Studies I–III we used the same sample retrieved from multiple registers. Data were combined from eight Finnish national registers, six of which concern benefits connected to long-term illnesses or disabilities, allowed by the Social Insurance Institution of Finland (SII), and two concern care provided by hospitals or social welfare. The SII registers concerned Child Disability Allowance (CDA), Disability Pension (DP), Disability Allowance (DA), Pensioners' Care Allowance (PCA), Funding of Rehabilitation (REH), and Preferential Refunding of Long-term Medication (MED). The other two were the Hospital Discharge Register (HOSP), and the Care Register for Social Care (CARE) kept by the National Research and Development Centre for Welfare and Health. Register sampling was based on unique personal identity codes (PICs). Every person in Finland has a PIC, which is used in every register to identify individuals. We generated a list of ID diagnoses (according to both ICD-9 and ICD-10) to be used as inclusion criteria (Appendix 1). Of the aetiological diagnoses, only those in which ID is typically present were included. *Table 9* gives an overview of the registers.

The search was carried out using data concerning the year 2000 in all registers except the Hospital Discharge Register. As a hospital stay may be a solitary event in a person's life, a 1-year search period may be too short to identify most cases. All persons with an ID diagnosis who had been in hospital during 1996–2000 and were still alive in 2000 were identified. Because the register started using the ICD-10 classification in 1996, a period of five years was used.



All persons fulfilling the inclusion criteria in any of the registers were compiled into one list. Double appearances were prevented by the unique social security code given to every Finnish citizen.

The process of how the benefits of CDA, DP, DA and REH are applied, and how the decisions for benefits are made in practice is illustrated in *Figure 13*.

Figure 12. Flow chart of the process for application of the following benefits: CDA, DP, DA, and REH.

Table 9. The benefit and service registers used in the multiple register survey.

A. Benefit registers of the Social Insurance Institution	
Child Disability Allowance (CDA) <ul style="list-style-type: none"> • Ages 0–15 years • Diagnosis recorded • Search from year 2000 	The SII pays CDA to support the care of a disabled or chronically ill 0–15-year-old child at home and to compensate for additional expenses and special arrangements. The allowance is paid regardless of the parents' or child's income or assets.
Disability Pension (DP) <ul style="list-style-type: none"> • Ages 16–64 years • Diagnosis recorded • Search from year 2000 	The DP, granted to individuals aged 16–64 years, provides compensation for working incapacity, i.e. a person's inability to engage in gainful employment, and is not intended to compensate for the actual illness or disability.
Disability Allowance (DA) <ul style="list-style-type: none"> • Ages 16–64 years • Diagnosis recorded • Search from year 2000 	The DA makes it easier for disabled persons of working age (16–64 years) to manage their ordinary everyday activities and to cope with their work and studies. It is granted to persons with an illness or injury that reduces their functional capacity for a period of at least 12 months. The allowance covers costs of a general handicap, the need for assistance, services, guidance or supervision, and additional expenses.
Pensioners' Care Allowance (PCA) <ul style="list-style-type: none"> • Ages 16+ years • Diagnosis recorded • Search from year 2000 	The PCA enables pension recipients with an illness or disability to live at home, supporting their home care and reimbursing recipients for extra costs incurred by illness or disability. Persons eligible for PCA are Finnish residents who are receiving either Disability Pension or old-age pension, aged 65 years or more.
Funding of Rehabilitation (REH) <ul style="list-style-type: none"> • Ages 0–64 years • Diagnosis recorded • Search from year 2000 	The SII grants funding for rehabilitation in conjunction with funding from other organizations. The special focus is the rehabilitation of severely handicapped 0–64-year-old persons. This register covers only the funding decisions made in the SII.
Preferential Refunding of Long-term Medication (MED) <ul style="list-style-type: none"> • Any age • No diagnosis recorded • Search from year 2000 	Patients suffering from certain serious and long-term diseases are entitled to a full (100%) refund of medication expenses ('preferential refund') for a person of any age. One of the accepted indications is 'perturbation of mind'. The presence of ID must be proven, but no diagnosis was coded in this register at that time.
B. Registers of Hospital Care and Care for the Intellectually Disabled, kept by the National Research and Development Centre for Welfare and Health (STAKES, nowadays THL)	
Hospital Discharge Register (HOSP) <ul style="list-style-type: none"> • Any age • Diagnosis recorded • Search from years 1996–2000 	For every visit to hospital, one to six diagnoses are coded in this register upon discharge (according to ICD-10). All persons with an ID diagnosis who had been in hospital during 1996–2000 and were still alive in 2000 were identified. As a hospital stay may be a solitary event in a person's life, a 1-year search period may be too short to identify most cases. Therefore, a period of 5 years was used. Because the register in its current form was dated from 1996 onwards, we could not go further backwards.
Care Register for Social Care (CARE) <ul style="list-style-type: none"> • Any age • No diagnosis recorded • Search from year 2000 	The register contains information about housing of persons with ID, gathered in two ways: upon discharge throughout the year (temporary visitors) and as a census of all inhabitants at the end of the year (temporary visitors at that time or permanent inhabitants). Both information-gathering methods are used in institutions and in housing with 24-h assistance, but only the annual census is used in less-well supported or independent accommodation for persons with ID.

For prevalence calculation total Finnish population data were received from the Social Insurance Institution.

Table 10 gives an overview of how the samples were retrieved from various registers. In each age group the most important register(s) giving the highest number of ID cases ("major register") were different. In each age group (0–15, 16–39, 40–64, 65+) one major register covered 59–81% of the total cases. However, every register contained unique information, adding unique cases to retrieval in the multiple register study (Table 11).

Table 10. Numbers of cases of ID traced from different registers, and proportions of cases that could be traced from each register of the total number in that age group in parentheses. Figures in columns are overlapping – totals are from original data. The register giving the largest capture ("main register") is in bold. CDA = Child Disability Allowance, DP = Disability Pension, DA = Disability Allowance, PCA = Pensioners' Care Allowance, REH = Funding of Rehabilitation, MED = Preferential Refunding of Long-term Medication, HOSP = Hospital Discharge Register, CARE = Care Register for Social Care.

	0–15 y	16–39 y	40–64 y	65+ y	Total
CDA	3238 (60%)	263 (2%)	0	0	3501
DP	0	9117 (80%)	13,613 (81%)	330 (11%)	23,060
DA	0	343 (3%)	17 (0%)	0	360
PCA	0	5933 (52%)	5803 (36%)	173 (6%)	11,909
REH	1921 (36%)	1316 (12%)	328 (2%)	1 (0%)	3566
MED	122 (2%)	1857 (16%)	5347 (33%)	1767 (59%)	9093
HOSP	2770 (52%)	1636 (14%)	1784 (11%)	823 (27%)	7013
CARE	1325 (25%)	3904 (34%)	4571 (28%)	529 (18%)	10,329
TOTAL	5346 (100%)	11,417 (100%)	16,295 (100%)	2995 (100%)	36,053

Table 11. Numbers of cases occurring only in the specific register, and proportion of unique cases in each register in different age groups. CDA = Child Disability Allowance, DP = Disability Pension, DA = Disability Allowance, PCA = Pensioners' Care Allowance, REH = Funding of Rehabilitation, MED = Preferential Refunding of Long-term Medication, HOSP = Hospital Discharge Register, CARE = Care Register for Social Care.

	0–15 y	16–39 y	40–64 y	65+ y	Total
CDA	935 (17%)	59 (1%)	0	0	994 (3%)
DP	0	2153 (19%)	5029 (31%)	164 (5%)	7346 (20%)
DA	0	101 (1%)	12 (0%)	0	113 (0%)
PCA	0	232 (2%)	281 (2%)	45 (2%)	558 (2%)
REH	207 (4%)	146 (1%)	37 (0%)	0	390 (1%)
MED	4 (0%)	84 (1%)	863 (5%)	1364 (46%)	2315 (6%)
HOSP	1031 (19%)	480 (4%)	644 (4%)	643 (21%)	2798 (8%)
CARE	410 (8%)	410 (4%)	291 (2%)	272 (9%)	1383 (4%)
TOTAL	2587 (48%)	3665 (32%)	7157 (44%)	2488 (83%)	15,897 (44%)

4.2 STUDY IV

After the multiple register study was performed, coverage of the HOSP register had developed – now covering all diagnosed conditions in specialised care³. This gave the possibility to focus more reliably on the epidemiology of ID during developmental age, from birth until 18 years⁴. Thus, in this study we used only the HOSP register.

Cases covering 1996–2017 were retrieved from the HOSP register. A similar inclusion list of diagnoses was used as in register sampling for Studies I–III. Cumulative prevalence was calculated for each birth cohort for each year. To diminish random variation, three birth cohorts were combined. This gave four succeeding cohorts, 1996–1998, 1999–2001, 2002–2004, 2005–2007. The increase of cumulative prevalence was compared between these four cohorts. In annual calculation a person was excluded after death by cross-checking the data with the Finnish death register (Population Register Centre), and data from Statistics Finland (Official Statistics of Finland) was used to calculate the prevalence of ID in the Finnish population for each year and age group.

The distribution by level of ID was calculated by using the latest diagnoses in the F7 group in ICD-10.

4.3 ETHICAL CONSIDERATIONS

Collating of the data from different registers was carried out by using unique personal identity codes, which were thereafter replaced with running numbers. Thus, the researcher was blind to the identities of the persons. The study was approved by the Ethics Committee of the National Research and Development Centre for Welfare and Health (STAKES).

4.4 STATISTICAL ANALYSES

In Study I prevalence estimates were calculated in four age groups (0–15, 16–39, 40–64, 65+) and both sexes, and in Study II in one-year age cohorts.

In Study I a capture-recapture analysis (Tilling 2001) was performed by grouping the registers in two, i.e. those from SII (A1–6) and those from STAKES (B1–2).

In Study III a corrected estimate was calculated for the population above 65 years of age, because in Study II an abrupt drop was noticed in the age-specific prevalence distribution at 65/66 years. At the age of 65, persons with ID who had previously received a disability pension with a medical diagnosis are reassigned to the old-age pension register, where no medical diagnosis is recorded. For this reason, they can no longer be traced through the main register and only partly via other registers. They are thus effectively lost as ID cases, and the result is the noticed sudden drop. Calculation of the corrected estimate was based on the age distribution of the general population, and the age-specific prevalence was allowed to decrease from the average level observed in five preceding age cohorts before the drop until

³ The name of the register was changed in 1994 to the Care Register of Health Care, but here it is still referred to by the previous name, Hospital Discharge Register. In the original paper of Study IV the actual name is used. The name of the upkeep organization was also changed from the National Research and Development Centre for Welfare and Health (STAKES) to the National Institute for Health and Welfare (THL)

⁴ The register was developed so that the oldest age cohorts with enhanced coverage were 17 years of age at the time of register sampling.

the observed average prevalence in the oldest age cohorts. Validation of the corrected estimate was based on another finding in Study II. At the same age (65/66 years) the proportion of persons with ID having the MED benefit (numerator) calculated as a percentage of all persons with ID (denominator) increased abruptly, which was due to an abruptly decreased total number of persons with ID (denominator in the quotient).

In all studies 95% confidence intervals (CIs) were calculated for the prevalence estimates according to Gardner and Altman (1989).

5 RESULTS

5.1 STUDIES I AND II

In Study I we explored the prevalence of ID in the whole population of Finland divided into four age groups. The mean prevalence estimate of ID for the whole population was 0.77% (95% CI 0.76–0.78) for males, 0.63% (95% CI 0.62–0.64) for females, and 0.70% (95% CI 0.69–0.70) for all. Large differences in ID prevalence rates were present between the four age groups (≤ 15 , 16–39, 40–64, ≥ 65 years) (see *Table 12*).

The capture-recapture analysis increased the total prevalence estimate to 0.87% (95% CI 0.86–0.88); the increase was largest in the youngest and oldest age groups, by 25% and 144%, respectively, and less in the middle groups, by 12–13%.

Table 12. Prevalence of ID calculated from combined registers. 95% confidence interval in brackets.

	0–15 y		16–39 y		40–64 y		65+ y		Total	
Males	0.61%	[0.59, 0.63]	0.79%	[0.77, 0.80]	0.96%	[0.94, 0.99]	0.39%	[0.36, 0.41]	0.77%	[0.76, 0.78]
Females	0.45%	[0.44, 0.47]	0.61%	[0.60, 0.63]	0.87%	[0.85, 0.89]	0.38%	[0.37, 0.40]	0.63%	[0.62, 0.64]
Total	0.53%	[0.52, 0.55]	0.70%	[0.69, 0.71]	0.92%	[0.90, 0.93]	0.38%	[0.37, 0.40]	0.70%	[0.69, 0.70]

If only the main register for each age group had been used (0–15 y CDA, 16–64 y DP, see *Table 10*), the prevalence estimates would have been much lower. Likewise, the extent of inclusion diagnoses had a great influence: narrow list (ICD-9: 311–314 and ICD-10: F70–79), broad list (narrow list and Down's syndrome), or full list (Appendix 1). (See *Table 13*.)

Table 13. The impact on prevalence estimate when using only the main register instead of collating multiple registers, or using just a narrow list (ICD-9: 311–314 and ICD-10: F70–79), a broad list (narrow list and Down's syndrome), or a full list (Appendix 1)

Age, years	Only main register		Multiple registers
	Narrow	Broad	Full
0–15	0.17%	0.25%	0.53%
16–39	0.41%	0.47%	0.70%
40–64	0.60%	0.64%	0.92%

The MED register yielded 2315 cases (6.4% of total) who did not appear in any other register with any diagnosis of ID. However, 999 of these (43%) appeared in one or several registers of SII with some other diagnoses of disability or long-term illness.

To acquire a deeper understanding of how the prevalence varies with age, calculations were performed in Study II for one-year age cohorts instead of larger age groups (see *Figure 13*). Age distribution figures showed peaks and troughs. Several observations could be made. The prevalence started increasing steadily from 0.20% in the first year of life until there was a relative peak in 10-year-olds with a prevalence rate of 0.74%, then markedly lower prevalence in later age cohorts during developmental years. In adulthood, increasing prevalence with age yielded the same level (0.74%) in the

age cohorts older than 33 years, with steadily increasing figures in older cohorts. The highest level, above 1%, was noted in age cohorts of 45–50 years, born in 1950–55. In age cohorts of 52–64 years (born in 1936–1948), the average prevalence was again on a lower level, 0.89%. At the age of 65/66 years there was an abrupt drop in prevalence rates, from 0.88% to 0.50%. The age-specific rate then diminished between ages 67–74 years, and remained on a stable level, 0.3%, with random effects becoming larger due to the diminishing absolute numbers. Thus, between 16 and 64 years the prevalence rose from ca. 0.6% to ca 0.9%. There were, however, several age cohorts (42–52 years, born in 1948–58) with remarkably higher prevalence, up to 1.04%. In the age group 16–64 years, 82.0% of the cases could be traced from the DP register.

At the pension age of 65 years there was a sudden drop in prevalence to 0.49% (95% CI 0.43–0.55). As the Disability Pension (DP) benefit ceases at this age, cases could no longer be traced from the DP register. The replacing pension, old-age pension, does not depend on diagnosis, and the previous diagnoses are thus lost.

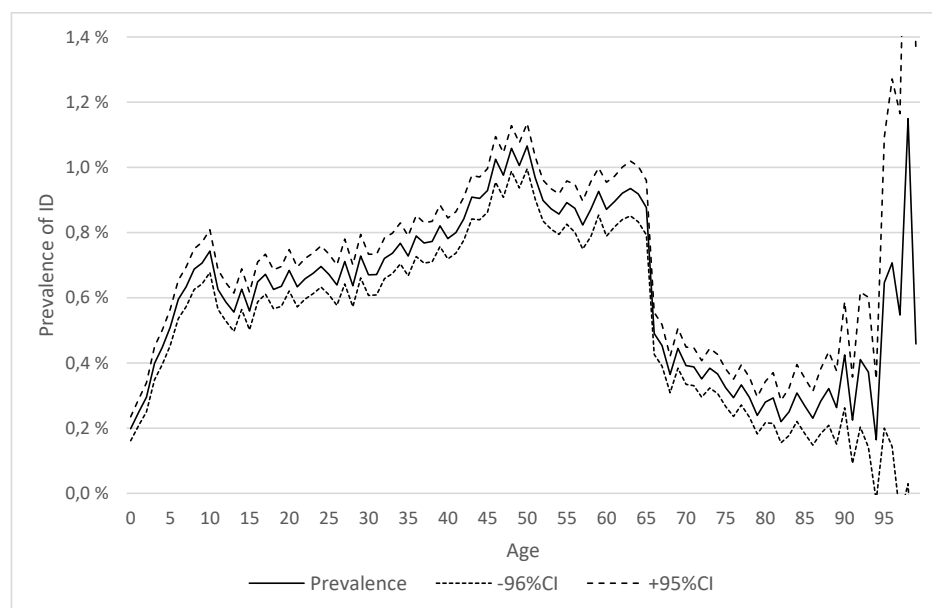


Figure 13. Age-specific prevalence distribution of ID, with 95% confidence intervals.

In gender distribution, there was a continuous trend from male preponderance in childhood to the opposite in late adulthood (Figure 14). This preponderance was much more marked than that in the general population, average 1.36 (0–18 y) vs. 1.04. The change to female preponderance took place some years later than in the general population.

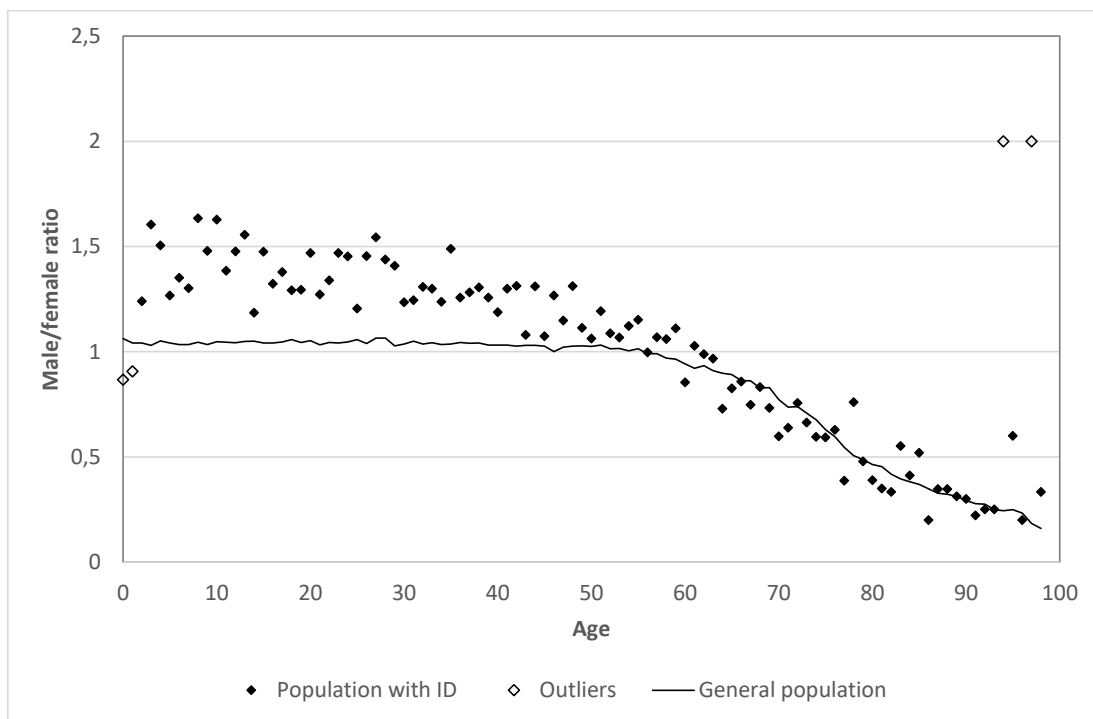


Figure 14. Ratio of number of males with ID to number of females with ID by age in one-year age cohorts in the multiple register study of ID. Outliers are marked differently.

To find possible explanations for the atypical features (increasing prevalence in adulthood, or, interpreted in the other way, decreasing by succeeding age cohorts and peak in prevalence among those born in 1948–1958) noted in the distribution (*Figure 13*), further analyses were made using the DP register. *Figure 15* shows at which age (decade) a decision on DP was made. In the oldest age cohorts (60–65 y), decisions had been made equally at different ages. In younger age cohorts, the decisions had been made mostly when the person had been under 31 years of age. Among recipients of DP, the decision was made at ≤ 20 years of age in 80% of the cases for age groups 16–47 years in 2000. As an example of the gradual increase of DP recipients in one age cohort, the accumulation is shown for those aged 50 in 2000 (*Figure 16*). After a rapid increase up to 80% at ages 16–21, new decisions for DP were then taken evenly throughout life. The same data can be seen from the viewpoint of the Social Insurance Institution, which makes the decisions (*Figure 17*). It must be noted that the statistics concern only persons alive in 2000, which means that in past years more decisions have been made concerning persons who are now deceased. For them, during the 1950's–60's the decisions were made mainly before age 31, and after that increasingly also to older persons. Major changes can also be seen in the incidence of decisions concerning the general population. Between 1955–1968 the annual incidence increased from ca. 4/10,000 to 14/10,000. Then during 1969–1986 the annual incidence decreased to ca. 6/10,000.

During 1989–1998 another increase was seen, up to more than 9/10,000, and thereafter a rapid decrease was seen during the analysed time.

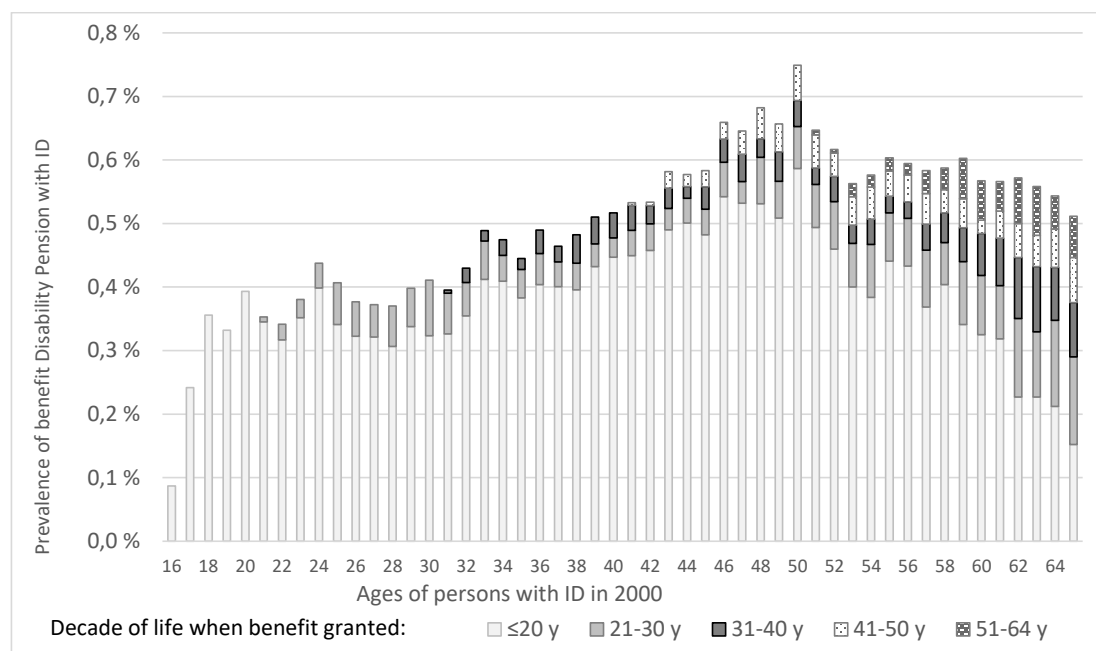


Figure 15. Decade of life when the decision for disability pension for ID was made for those alive in 2000, plotted by age in 2000.

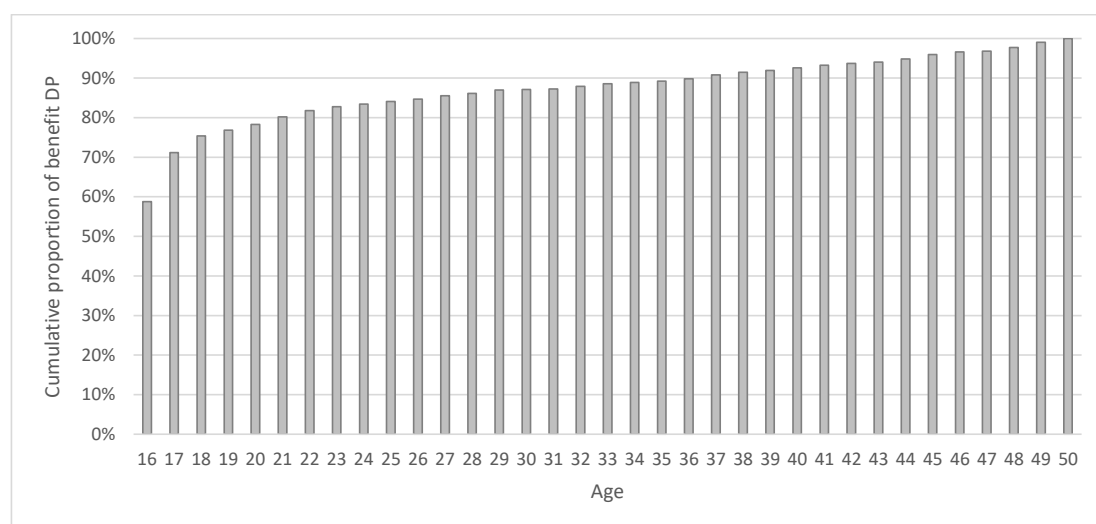


Figure 16. Cumulative proportion of Disability Pension benefit by age in the 50-year-old subgroup of the multiple register study.

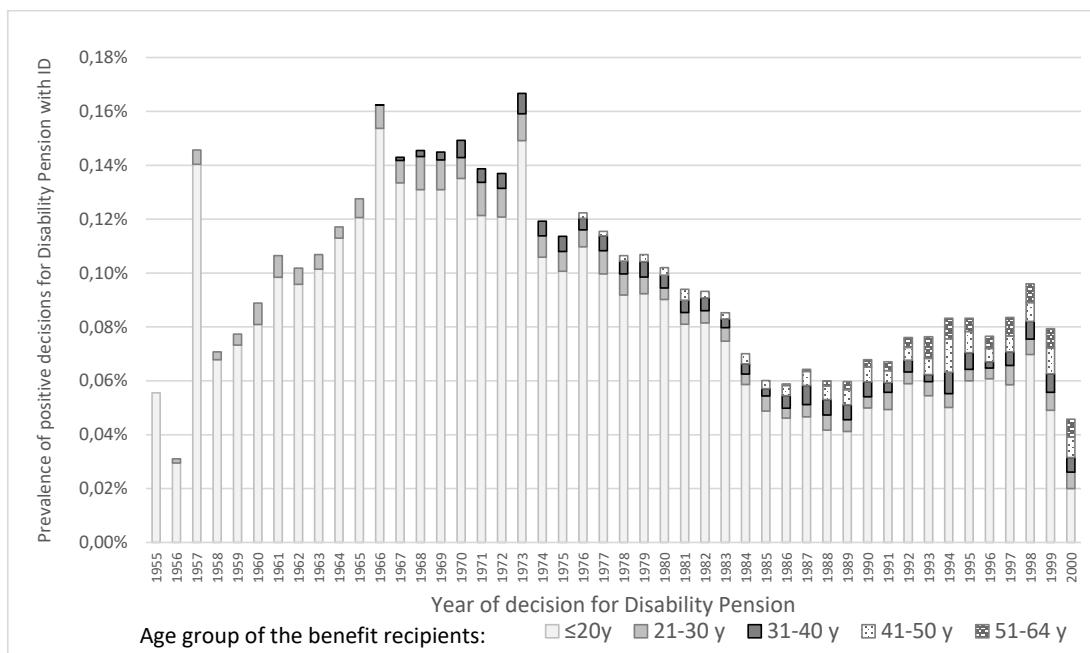


Figure 17. Annual decisions on disability pension after diagnoses of ID in different age groups; persons alive in 2000; administrative prevalence of ID calculated as the proportion of the current population at that time.

5.2 STUDY III

This study was performed to explore the deviant finding in the age-specific prevalence distribution at age 65 years in Studies I and II. We made a correction to smoothen the sudden drop.

The multiple register study of ID revealed an average prevalence of 0.38% (95% CI 0.37–0.40) among 65+-year-olds. At the age of 65/66, a sudden drop in the prevalence of ID was recorded, from 0.84%/0.91% to 0.49%/0.49% (men/women). The average prevalence rates in the five preceding age cohorts prior to the drop were 0.90% for men and 0.91% for women. The average prevalence observed in the oldest age groups (80+ years) was 0.30%. In correction of the estimate, age-specific prevalence was allowed to decrease evenly from 0.90% to 0.30%, calculated from the general population in year 2000, separately for females and males. After that the new prevalence estimate for the whole age group of 65+ years was calculated.

The graph of the prevalence distribution after correction is shown in *Figure 18*. After correction the average estimate for the age group of 65+ years was 0.75%, nearly doubling the estimate according to the registers. If in reality the increased mortality rate would cause the prevalence to drop faster among 75+-year-olds, this would not substantially influence the new total prevalence estimate of all old ages combined, since 63% of the increase in the prevalence estimate was connected to 66- to 75-year-olds. If the correction were to be applied only to this age span, the total prevalence among 65- to 100-year-olds would still be 0.61%.

In order to use the proportion distribution (those persons with ID having the benefit of Preferential Refunding of Long-term Medication of all persons with ID) for validation of the previous corrected

prevalence estimate, we had to check that the abrupt increase at 65/66 years was not connected to an increased allowance of the benefit at retirement age. Of the 66- to 70-year-olds in 2000 in the register sample, 87% had been granted the benefit over 10 years earlier. Thus, the abrupt increase was due to decreased total number, the denominator in the quotient. Correction of the age-specific prevalence estimate smoothed the curve at 65/66 years.

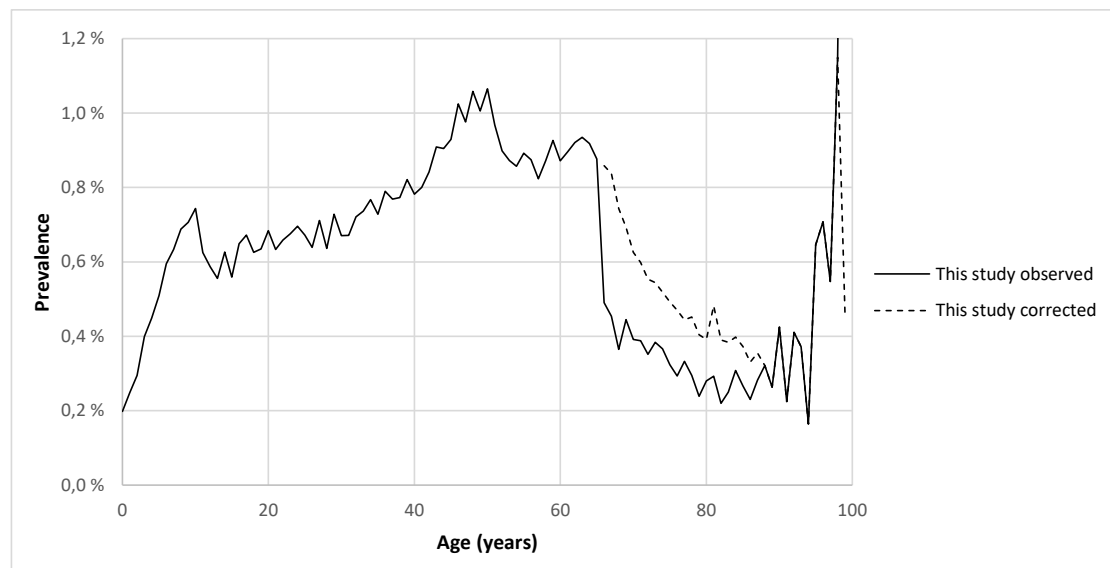


Figure 18. Age-specific prevalence distribution of ID before (solid line) and after (broken line) correction.

5.3 STUDY IV

In this study we calculated prevalence estimates of ID among children and adolescents in several succeeding birth cohorts (born 1996–2007) at different ages using the Hospital Discharge Register (HOSP) covering 1996 to 2017.

Soon after birth the prevalence was ca. 0.2%, and the cumulative prevalence increased steadily by age (Figure 19). The 1996 birth cohort could be followed up to age 17.5 years – the cumulative prevalence reached 1.19%. The increase with age occurred at a faster rate in the later-born age cohorts (1997–2007), but the final cumulative prevalence before adulthood could not yet be seen in 2017.

New cases with ID inclusion diagnoses were registered steadily throughout the developmental years (Figure 19). There was a somewhat greater incidence between the ages of four and seven. Among those born in 1996 and followed up until the age of 17.5 years, the prevalence reached 1.19% (95% CI 1.11–1.28) for all, 1.42% (95% CI 1.29–1.55) for boys, and 0.95% (95% CI 0.84–1.06) for girls. For those born three years later, in 1999, a comparable prevalence rate (1.21%) was reached earlier, by the age of 14.5 years. When comparing the four succeeding three-year birth cohorts (1996–1998, 1999–2001, 2002–2004, and 2005–2007), the cumulative prevalence at 4.5 years was roughly the same (0.46–0.49%). Beyond that age, however, ID diagnoses were recorded at an earlier age in later-born age cohorts. By the age of 6.5 years, the cumulative prevalence of ID was 0.66% (95% CI 0.62–0.70) for those born in 1996–1998, but was 0.80% (95% CI 0.76–0.84) for those born in 2005–2007.

The difference between cumulative prevalence based on a register search throughout developmental years and point prevalence based on a one-year search was large. When the search was performed in connection with one year only, the prevalence figure was on average 0.30% (maximum 0.58%) depending on the age and year of concerned.

The impact of mortality on the final cumulative prevalence was relatively small. Of all cases identified during the study period, 4.6% had died. In the oldest birth cohort (born in 1996) and followed up until the age of 17.5 the death rate was 4.1%. Therefore, without deaths the final cumulative prevalence would have been 1.23% instead of the observed 1.19%.

Both means of including diagnoses in registers – by various aetiological diagnoses and by level of ID (F79–F79) – brought subjects into the register catchment. By the end of the follow-up period, 67.2% had received an F7 diagnosis, 42.2% had an aetiological inclusion diagnosis, and 9.4% had both an F7 and an aetiological diagnosis. Among those with only an aetiological diagnosis and no F7 diagnosis, Down’s syndrome was the most frequent diagnosis. Only 8.5% of people with Down’s syndrome held a parallel F7 diagnosis.

The distribution of developmental level by F7 diagnoses was calculated for those born in 1996–1998 and followed up to the age of 15.5–17.5 years. In this group, 75% had an F7 diagnosis. Of these, 79% had a defined level of ID; for others it was unspecified (F78–F79). The proportion of sID was 28% among those whose level was diagnosed. The population prevalence of diagnosed sID was 0.20% ($1.19\% \times 0.75 \times 0.79 \times 0.28$). If the same distribution of levels of ID was also true concerning those without a defined level diagnosis, the prevalence estimate of sID would be 0.33% ($1.19\% \times 0.28$).

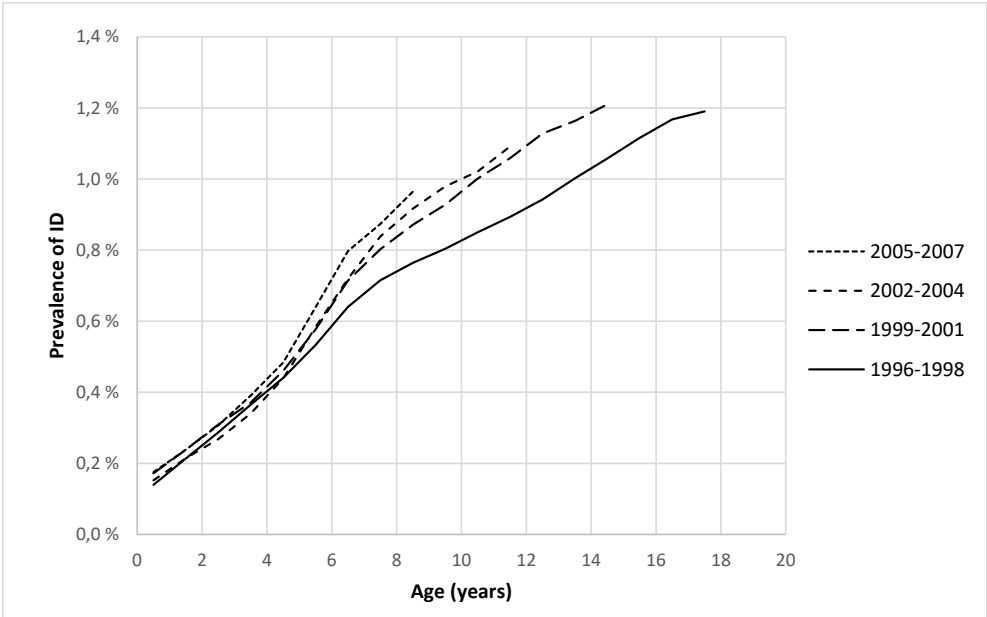


Figure 19. Cumulative prevalence of ID at different ages in four succeeding age cohorts, 1996–1998, 1999–2001, 2002–2004, and 2005–2007.

6 DISCUSSION

Average prevalence estimates for ID were generated for different age groups from the multiple register data, 0.53% for 0–15-year-olds, 0.70% for 16–39-year-olds, 0.92% for 40–64-year-olds, and 0.38% for 65+ year-olds. When exploring these rough estimates in more detail, the prevalence calculated by using one-year age cohorts revealed interesting differences with age both within and between the four age groups. From age 16 to 64, the age-specific prevalence increased continuously, with an extra peak at 42–52 years of age. An abrupt drop was noticed at 65/66 years of age. In Study III, calculation was performed to correct the estimate in the oldest age group – the corrected estimate of 0.75% was almost double that of the one yielded by registers. In Study IV, new cumulative prevalence estimates from the HOSP register were calculated for under 18-year-olds. At the brink of adulthood, a prevalence of 1.19% was reached among those born in 1996. In later-born age cohorts the cumulative prevalence increased faster. For those born three years later, a prevalence of 1.21% was already reached at 14.5 years of age, but in this study the final prevalence concerning them could not yet be seen in 2017.

6.1 STRENGTHS AND LIMITATIONS

The method of looking at one-year age cohorts provided plenty of information (Studies II–IV). It gave a deeper view of the population with ID and helped to generate new questions for further studies. Using a broad list of inclusion diagnoses gave a greater coverage than when limited only to ICD-10 group F70–79. The possibility to combine registers based on personal identity codes, thus preventing double counts, increased coverage and reliability. Finland's strength is to have national registers of good quality (Gissler and Haukka 2004, Lampi et al. 2010, Sund 2012, Leivonen et al. 2014). A practical strength is the fact that information is collected in registers along with normal social and health services all the time, which provides easily accessible time series for research purposes.

Concerning coverage, a major weakness is that those in clinical practice do not regularly use ICD diagnoses of ID (ICD-10: F70–F79) alongside aetiological diagnoses. In this study we could add to the inclusion list of diagnoses only those where ID is regularly present, but those where it may only occasionally be part of the clinical picture would also have needed the F7-group diagnosis.

Coverage of the registers concerning social benefits is limited, since not all with a diagnosis of ID are eligible for benefits. At any stage of the process ending with coding in a register, there are losses of cases (see *Figure 12*), and even combining such registers does not achieve complete coverage. In contrast, the Hospital Discharge Register contains information on diagnostic assessments irrespective of whether social benefits or services are granted. Having this national register available is a major strength.

Some of the registers used are cross-sectional by nature, such as the Hospital Discharge Register. To acquire the most comprehensive picture from such registers, the study should be longitudinal, continuing all the time. This was only possible in Study IV. On the other hand, some of the registers hold information on permanent or long-term benefits for ID. One limitation is that in Studies I–III the study sample was fairly old (year 2000). The sample was retrieved for Study I, and to allow further explorations the same sample was used for Studies II and III. Furthermore, it allowed us to explore historical cohorts born in 1948–1958, and to compare them with prior Finnish cohorts (Ruoppila 1966).

It is not possible to know exactly what methods have been used in diagnostics. A study that covers decisions made over several decades and in different clinical units will always be limited in reliability. When diagnostic systems change, it is not possible to know how the new protocols have been implemented in units which report to the registers. The registers, even when collated, do not include all relevant information.

A medical diagnosis of ID does not tell us enough about coping and need for support, even in cases where aetiology, level of ID, and comorbidities have been given. Registers of social benefits (Invalidity Pension, Child Disability Allowance) inform us that the applicant has been assessed as needing a certain level of extra support to be eligible for the benefit. However, needs of support are not known for those who are not accepted to receive the benefit in question. Thus, the validity of data for estimating the need for services is limited, even when prevalence estimates are exact and uniform.

6.2 COMPARISON WITH PREVIOUS STUDIES

The estimate of prevalence of total ID for 0–17-year-olds, 1.19%, is slightly higher than the median of 1.10% in register-based studies and 1.03% in population sample studies (see review). It is higher or at the same level as in previous Finnish studies concerning developmental years, except for that by Kääriäinen (1987): Helsinki 1955 birth cohort study, 0.91% / 14 y (Amnell 1974), Finland-in-Miniature study 1962, 0.94% / 15–19 y (Ruoppila 1966), the Northern Finland 1966 Birth Cohort study, 1.19% / 14 y (Rantakallio and von Wendt 1986), the District of Kuopio study, 1.38% / 8–9 y (Kääriäinen 1987), and the Northern Finland 1985/86 Birth cohort study, 1.12% / 11.5 y (Heikura et al. 2003).

The estimate of sID in the present study was uncertain as a result of the large proportion (40%) with no or non-defined diagnosis of level of ID. The observed estimate, 0.20%, is well under the median mentioned above in the review, 0.42%. If the observed figure is extrapolated to the non-defined, even that estimate, 0.33%, would be slightly under the median. This extrapolation can be considered justified, because there is less uncertainty concerning severe forms of ID during follow-up, and there is a readiness to state a specific diagnosis. In comparison, in the Helsinki 1955 birth cohort study the prevalence estimate for sID was 0.39% / 14 y (Amnell 1974), in the Finland-in-Miniature study 1962, 0.50% / 15–19 y (Ruoppila 1966), in the Northern Finland 1966 Birth Cohort study, 0.63% / 14 y (Rantakallio and von Wendt 1986), in the District of Kuopio Study, 0.63% / 8–9 y (Kääriäinen 1987), and in the Northern Finland 1985/86 Birth cohort study, 0.38% / 11.5 y (Heikura et al. 2003). There may be a decreasing trend in prevalence of sID over time, which may be confounded by regional differences.

In line with the results of previous studies in which the prevalence of ID has been given for various child/adolescent age groups (under 18 years), prevalence increased with age. In addition, the rate of increase by age (the slope of the rising line over time) is becoming steeper in comparison with the results of most studies (*Figure 20*). A similar and even faster change has been noted in connection with several neuropsychiatric disorders (autistic spectrum disorder, hyperkinetic disorder, obsessive-compulsive disorder, and Tourette's syndrome) in Denmark, Finland, Sweden, and to some degree in Western Australia (Atladottir et al. 2015). The authors attribute the changes to shared non-aetiological factors, such as improved service availability, broadening of the diagnostic criteria, and increased awareness of neuropsychiatric difficulties in both the lay and professional communities. They also point to the possible previously unmet need for services, changes in the healthcare service systems, and also more frequent non-intact family structures, which may increase the extent of help-seeking behaviour.

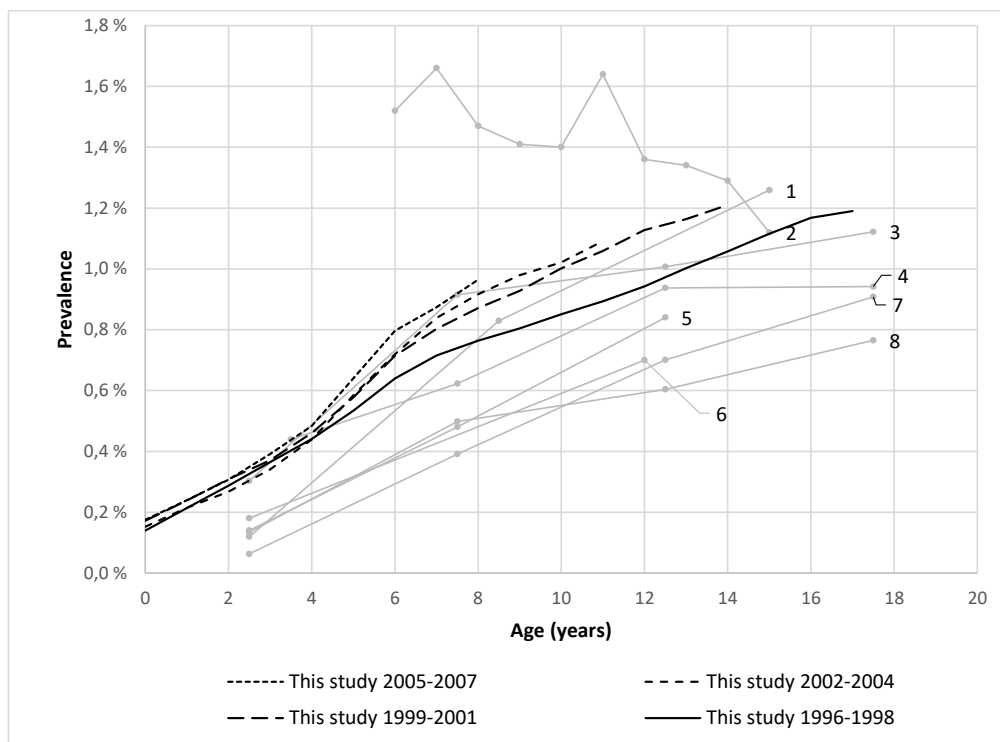


Figure 20. Comparison of Study I cumulative prevalence of ID with data from previous studies. Cumulative prevalence by age in four different birth cohorts. Studies are: 1) Fishbach and Hull 1982, 2) Leonard et al. 2003, 3) Doyle and Carew 2016, 4) Ruoppila 1966, 5) Brask 1972, 6) van Schrojenstein Lantman-de Valk et al. 2006, 7) Díaz-Fernández 1988, and 8) Yen et al. 2013.

Our estimate of the prevalence of ID in adults is very different from those in previous studies, when the age-specific distribution is considered (see Figure 21). In the younger part, 16–39-year-olds, the group estimate of 0.70% is roughly at the same level as in prior studies (see Table 5). The change in prevalence at later ages, however, is in contrast with those in most previous studies. The age-specific prevalence increased during adulthood instead of decreasing, and the group estimate of 0.92% for the 40–64-year-olds is high. There is only one other study in which the same observation was made (Ruoppila 1966), this being a population sample study.

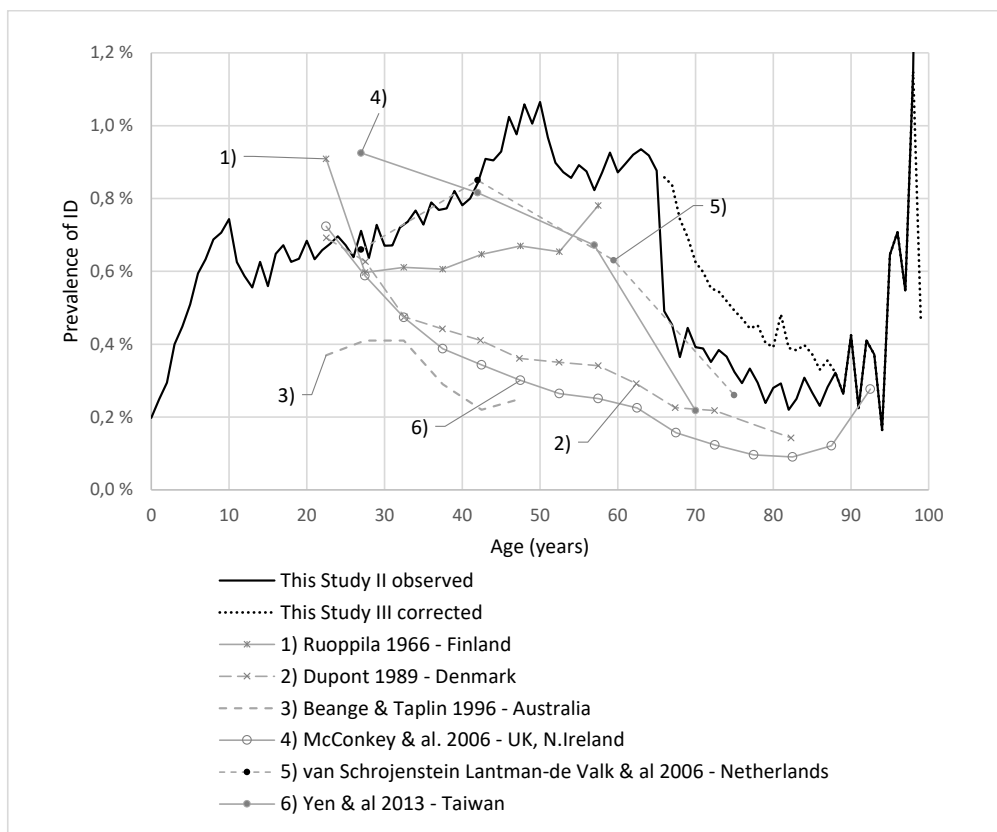


Figure 21. Age-specific prevalence distribution of ID in adulthood compared with previous studies.

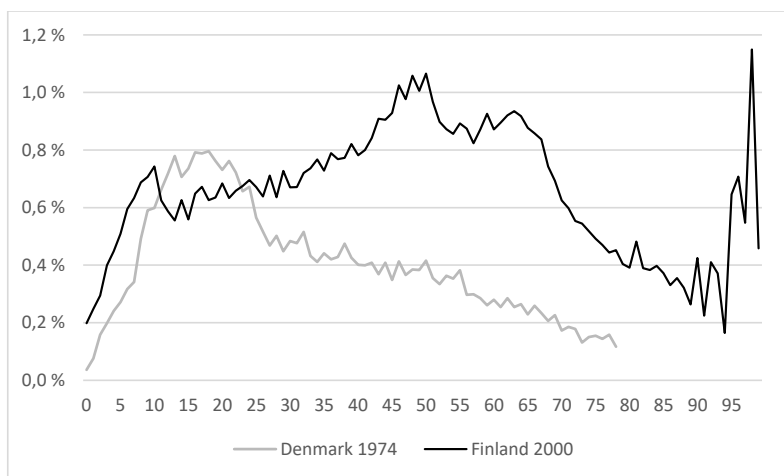


Figure 22. Comparison of current study results with those in Denmark in 1974 (Dupont 1975).

Concerning older persons, the difference between the current estimate of prevalence of ID and those in previous studies was most significant. Even without the correction, the estimate of 0.38% is higher than in most of the reviewed studies (*Table 6*). However, Tyrer et al. (2007) reported a prevalence rate of 0.35% for sID only. Our corrected estimate of 0.75% is much higher than in any of the previous studies (*Table 6*).

In *Figure 22*, a comparison of ID prevalence rates is made with the only other study found where the prevalence of ID is shown in one-year age cohorts (Dupont 1975).

The male-female gender ratio is in line with previous reports (Maulik et al. 2011). However, no previous study could be found where the gender ratio has been explored in one-year age cohorts. Our data enabled us to notice how the much higher preponderance of males than in the general population gradually diminished to turn to female preponderance some years later.

6.3 SIGNIFICANCE OF THE FINDINGS

For the background of discussion of the findings, a theoretical age-specific prevalence distribution can be constructed according to the following premises:

- Based on the definition of ID, the condition should be diagnosed at developmental age, that is, before the age of 18 years, up to which age the prevalence should reach its peak level.
- Intellectual disability can be diagnosed immediately after birth only in a minority of cases (e.g. Down's syndrome and other chromosomal or congenital conditions). Most diagnoses are, therefore, made later in childhood and adolescence.
- Mortality is higher among persons with ID than in the general population. A higher mortality rate in early/young childhood is a result of serious somatic disorders. In that period mortality has a downward influence on ID incidence. After that, mortality is somewhat higher than in the general population until becoming more marked in middle and older age, especially in severe ID, and in Down's syndrome, where Alzheimer's dementia shortens life expectancy remarkably.

If the prevalence rate of ID at 18 years is assumed to be 1%, for example, these premises would result in a distribution curve like that in *Figure 23*. The steepness of increasing and decreasing parts will depend on the incidence and mortality parameters in the study population, but it should not change the general configuration of the curve. Any changes in the observed distribution that are markedly different from those expected warrant an explanation.

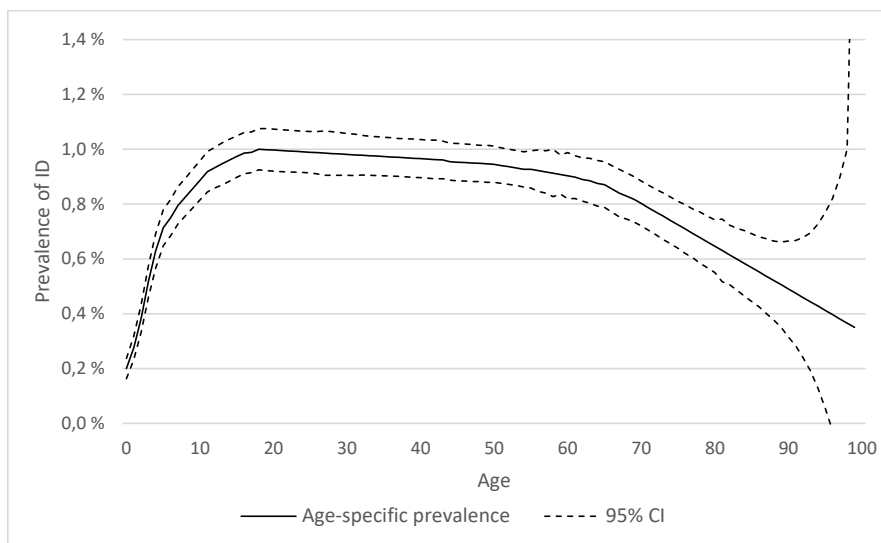


Figure 23. Theoretical age-specific prevalence distribution of ID in a population whose structure matches that of Finland in the year 2000.

As potential explanatory factors for observed differences in comparison with the theoretical distribution, the following may exert influence variably from time to time: changes in incidence of ID, activity and timing of clinical investigations for ID, varying diagnostic practices concerning ID, activity in and criteria for arranging services for ID persons with case registration, survival of persons with ID compared with those in the general population (in connection with diseases, wars, etc.).

The prevalence estimates from all four studies are combined in Figure 24. The combined results can be analysed according to the presented premises, by age groups and from several viewpoints.

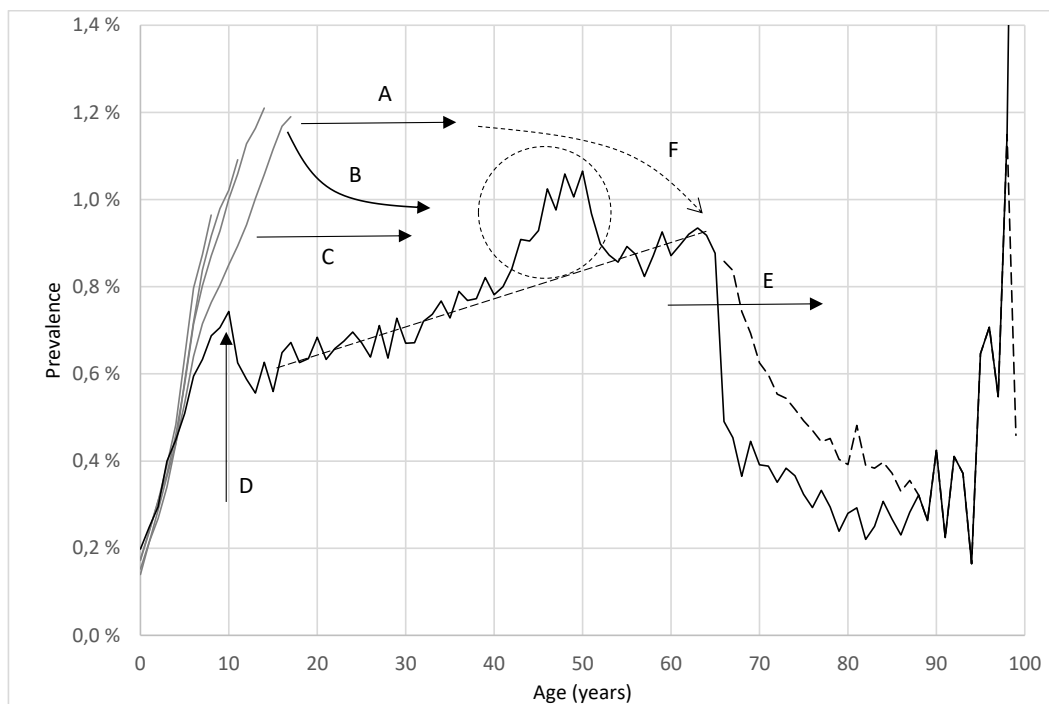


Figure 24. Results of Studies II–IV combined. The solid bold line is the distribution from Study II, the broken bold line is the corrected distribution from Study III. The four grey lines are the prevalence distributions of succeeding age cohorts from Study IV. The following remarks are discussed in the text. Arrows marked A, B and C denote different hypothetical continuations of the age-specific prevalence figures. D points to the peak prevalence in childhood in the multiple register study. Arrow E refers to persons surviving beyond pension age. Arrow F refers to the decrease in prevalence due to increased mortality. The thin broken line is the trend line for prevalence between ages 16–64. The dotted circle marks the exceptionally high prevalence in the age cohorts of 43–52 years.

6.3.1 Childhood/adolescence

Cumulative prevalence estimates in childhood are based on Studies II and IV. Study II yielded the highest prevalence (0.74%) at the age of 10 years using multiple registers including the Hospital Discharge Register (HOSP) for years 1996–2000, whereas Study IV, where we used only the HOSP register but a larger span of years (1996–2017) yielded the highest prevalence (1.21%) at 14.5 years of age for those born in 1999. In the multiple register study (Study II), half of the cases of 0–15-year-olds were traced from the HOSP register, which yielded all cases in Study IV. In Study II we used HOSP register data covering only 1996–2000, because register data was not available for earlier years. If a larger span of years had been available for Study II, the increasing curve might have continued from point D in Figure 24 upwards, as in Study IV, but probably less steeply. Extrapolating the line might have reached a prevalence estimate of ca. 0.9% at the age of 18.

There may be several reasons for the more rapid increase in the cumulative prevalence of ID by age in succeeding birth cohorts. More comprehensive screening and arranging of diagnostic assessments in specialist care is a very probable explanation – the change with age cohorts occurred most prominently after the age of four, when nationwide screening is performed in child guidance centres (Valtonen et al.

2004). Such screenings covered over 84% of all child guidance centres in 2007 (Hakulinen-Viitanen et al. 2008). There is also the possibility of an increasing incidence of ID, e.g. due to some particular aetiological diagnoses which would be reflected in an increase of register-based prevalence. The general trend of decreasing infant mortality may be reflected in the incidence of ID at later ages during development. Moreover, the quality and coverage of the HOSP register may have increased during the study period, especially since the earliest years.

Compared with old Finnish surveys, better epidemiological methods have also included persons with mID, and the estimate for the total prevalence of ID has increased. The estimate built on the Finland-in-Miniature study (Ruoppila 1966), the two Northern Finland Cohort Studies (Heikura et al. 2003) and those by Kääriäinen (1987) and Amnell (1974), has been around 1–1.3% for children and adolescents. That figure is in line with present study estimate.

6.3.2 Adulthood

As noted above, the prevalence rate of ID in adolescents entering adulthood may be as high as 1.2%, whereas the average in succeeding years (16–39 years) was 0.70%. How can the large discrepancy between the two studies be understood? The prevalence estimate for 17-year-olds in 2013 was almost twice as large as in the multiple register study in 2000. The time difference can only partly explain it – that would mean a large increase in prevalence in early childhood in age cohorts born in 1996–2017.

Where is the line continuing in adulthood (*Figure 24*, A, B, or C)? Does the prevalence line continue at 1.2% for some time (A) until aging will make it turn downwards due to higher mortality than in the general population? Or is the cumulative prevalence estimate too high throughout childhood and adolescence – not laying enough emphasis on adaptive skills in diagnosis (C)? Or does the curve bend downwards after childhood, when practical skills better compensate for difficulties in academic skills (B)?

There are two Finnish prospective studies with long follow-up periods that can shed light on the question of later coping and need for services among those who have been diagnosed as having ID during their developmental years.

A follow-up study of the Northern Finland 1966 Birth Cohort was undertaken in 2000, at the age of 34 years (Taanila et al. 2005). The prevalence of ID was 1.11% in 2000. All persons with sID and only 70% of those with mID were receiving Disability Pension. Further, coping of those not receiving DP was evaluated by working history and unemployment compared with a reference group (not diagnosed with ID in the cohort). Their unemployment rate was 56% compared with 12% in the reference group, and the median duration of unemployment was 138 days per year compared with 10 with the reference group. These statistics can be interpreted as meaning that a major proportion of those with mID have not been able to develop adaptive skills to cope well in the society in which they live. The findings of Taanila et al. (2005) suggest that the prevalence of ID around the age of 34 years cannot be reliably estimated by Disability Pension statistics. This finding raises the question of whether or not there is a so-called hidden population with ID (Whitaker 2013).

A follow-up study of the Finland-in-Miniature sample was performed in 1998, 35 years after the initial study (Ruoppila et al. 2003). In this study the prevalence of ID was originally 0.91–0.94% among 10–34-year-olds, who were 45–59-year-olds in 1998. Of those diagnosed in 1962 to have ID, only 43–48% were receiving Disability Pension in 1998, depending on the age cohort.

These two comparisons suggest underestimation of ID prevalence in the multiple register study concerning an adult population.

Why was the prevalence estimate increasing steadily with age in the multiple register study between the ages of 16–64 (born between 1936–1984) (*Figure 24*, trend line)? Or – expressed in the opposite way – why was the prevalence rate decreasing with advancing birth cohorts? Because 80% of the cases in the multiple register study were detected in the Disability Pension register in the age group of 16–64 years, one explanation could be gradually increasing allowances of the benefit with age, due to increasing problems based on ID. This is partly true. The DP benefit has been mostly granted at ≤ 20 years of age, and thereafter evenly by age (see *Figure 15*). However, the prevalence of those who were granted the DP benefit at ≤ 20 years of age has decreased by age cohort from 0.59% among 50-year-olds to 0.31% among 28-year-olds (see *Figure 15*). Does it then reflect a slowly decreasing incidence of ID in childhood/adolescence over the past decades? This might be a natural consequence of increasing health and nutrition, and especially prevention of those childhood diseases that hinder psychological development. However, the findings of Study IV, the much higher cumulative prevalence at the brink of adulthood (1.19%) challenge this hypothesis. For some reason, there have been drastic changes concerning the provision of disability pension by the SII, especially seen in the decreasing trend between 1966–1989 (*Figure 17*). Does this reflect the supposed decrease in the incidence of ID? Or has there been a decrease at some stage of the application process for the DP benefit (see *Figure 12*)? For example, have the criteria for granting the application become stricter? It was not possible to investigate these hypotheses in this study.

Why is there the exceptionally high peak in prevalence at 42–52 years (*Figure 24*, circle)? Those persons were born in 1948–1958, mostly after the so-called baby-boom generation (in Finland born 1945–1950). Going backwards in time, in the Finland-in-Miniature study, in 1962 those persons were 4–14-year-olds. At that time the prevalence was not higher in that age group compared with older age groups (Ruoppila 1966). If the finding reflects a period of exceptional incidence (medical aetiology), there would have been only some years of childhood/adolescence (14–17 y) in which more diagnoses in the healthcare system would have had an impact on the prevalence figure. In comparison of this group with similar age groups before and after (all covering 11 one-year age cohorts), there were no differences in the proportions of epilepsy or cerebral palsy to suggest severe medical aetiologies (unpublished analysis of the data). Other analyses could not be made from the data. The other possibility is that there has been a period of different criteria in making decisions on DP, i.e. relatively more among these cohorts than those before and after. However, from the data available, the age group of 42–52-year-olds does not differ from older or younger ones concerning the age when a positive decision regarding DP was made (see *Figure 15*), and neither have there been any marked changes in the activity of making those decisions in the Social Insurance Institution (*Figure 17*) concerning just this age group, although great changes in activity can be noted in the long term. Thus, the reason for the exceptionally high prevalence in this age group remains unknown.

6.3.3 Old age

The estimated prevalence of ID in old age in the multiple register study was 0.38% for the whole age group above 65 years. The estimate was then mathematically corrected in Study III, and the new estimate was 0.75%. However, the abrupt drop with correction at the age of 65/66 does not look correct (*Figure 24*). The prevalence profile would be more acceptable if the line continued from the level attained at the end of developmental age (F in *Figure 24*) according to the theoretical assumptions

(Figure 23). Another remarkable aspect concerns aging, i.e. persons in middle age moving to old age, from age cohorts with an exceptionally high prevalence of ID (Figure 24, E). One can suppose that even now the ID prevalence estimate in old age might be much higher than at the time of register sampling, in 2000.

Concerning older people, there were no previous Finnish studies on the prevalence of ID, but an analysis on the standardized mortality rate hinted at increasing prevalence (Patja et al. 2000). The few international studies have given lower estimates (see Table 6), but some investigators have also noted increased longevity (Carter and Jancar 1983, Janicki et al. 1999). This study truly showed the possibility of a hidden population of elderly persons with ID.

6.3.4 Gender ratio

The male-female ratio was much higher in childhood and adolescence, average 1.36, than in the general population, average 1.04. The higher incidence of ID among males has been partly explained by the frequency of early complications and X chromosome-linked syndromes (Harris 2006). However, this preponderance became reversed at the ages of 60–65y, being then similar to that in the general population. The greater longevity of females in the general population seems to be exaggerated in the population with ID, either via the same or different mechanisms.

6.3.5 Total estimate

From the prevalence estimates in different age groups, we can calculate a composite estimate for the entire population with ID based on the current study (Table 14). The assumptions are: prevalence increases from 0.2% to 1.2% between 0–10 years, then stays at the level of 1.2% between 11–17 years, decreases steadily from 1.2% to 1.0% between 18–64 years, and decreases rapidly from 1.0% to 0.3% between 65–100 years. As has been stressed in previous discussion, there are many uncertainties in this estimate. Because the uncertainty is more towards under-coverage, the estimate represents a minimum.

Table 14. Estimates for the total population with ID in Finland calculated for the population structure in 2017.

Age group	Average prevalence	Size of population 2017	Estimated number with ID	
0–10 y	0.6%	649,766	3900	(3899)
11–17 y	1.2%	416,495	5000	(4998)
18–64 y	1.1%	3,267,551	35,900	(35,943)
65+ y	0.75%	1,179,318	8800	(8845)
TOTAL	0.97%	5,513,310	53,700	(53,685)

7 CONCLUSIONS

The results of these multiple register studies suggest a higher estimated prevalence of ID than in most prior register-based studies in Finland, the estimate being close to that found in birth-cohort studies. However, persons with ID cannot be found effectively in registers after the age of 65 years and consequently there was an apparently large drop in prevalence of ID at this age. However, calculating a correction, the prevalence of old persons with ID was estimated to be much higher than previously thought.

The prevalence given in one-year age cohorts across the whole age span gave a qualitatively different picture and higher prevalence rates than more robust grouping by age. Inconsistencies in the age-specific prevalence distribution, together with other previous findings, hint at the possibility of so-called hidden disability, i.e. people with difficulties in coping, but not recognized by the services.

The cumulative prevalence of ID increases steadily throughout developmental years. Cross-sectional studies at any age do not give a full picture.

Different registers complement each other. Some reflected diagnostic activities, others, service delivery.

The prevalence distribution at all ages reflects in many ways the history of social and health care, both positive (development) and negative influences (epidemics, depression).

Continuous monitoring of the epidemiology of ID by one-year age cohorts via the most informative registers seems to be both useful and practical.

8 IMPLICATIONS FOR THE FUTURE

Some of the questions that were left open in this study can be answered with further register-based studies. The question of how the cumulative prevalence of ID will increase by age during childhood and adolescence, and will the final figure at 18 years be higher than before, may be investigated with the aid of the Discharge Register.

The fate in later life of those diagnosed with ID before 18 years is an important topic to be explored. A marked gap remained between the final cumulative prevalence estimate at the end of the developmental period (Study IV: 1.2%) and the estimate in early adulthood (Study II: 0.7%). These two estimates were derived from different data, but the topic needs further study. Is the present prevalence in early adulthood higher too? Is it possible and safe to assume that the persons with ID in childhood and adolescence will cope well enough to disappear from the service registers? Do they have enough support? Do they belong to the population about whom several clinicians and/or researchers have been worried (Peltopuro et al. 2014) – the "hidden ID population" (Whitaker 2013) or "lost generation" (President's Committee on Mental Retardation 1999)? Thus, new long-term studies are needed to find out whether and how persons with mild ID during their developmental years might have adaptive problems at an age later than 18, and how our society responds to their needs. Combining employment and education statistics to those of health and social care may provide more important information.

The size of the elderly population with ID can be estimated by way of longitudinal retrospective register sampling – how those who were receiving Disability Pension before age 66 survived in later life.

In the Introduction it was noted that reliable, up-to-date, and detailed epidemiological information is needed to organize, run, and reform service systems for persons with ID. In Finland, the current major reforms in social and health care have an emphasis on the need of a knowledge base. It has not been determined how exact this information should be for individually calculated capitation-based payment, or how sufficient are national and regional prevalence figures (Collings et al. 2016). Up to now there have been three main methods in different countries to monitor the appropriateness of services for those who are in need: 1) keeping a register of all those who need services, and collecting all necessary information, 2) making estimates from information that is collected from general registers, and 3) performing cross-sectional studies at times to evaluate the situation.

The aim of this study was not the profiling of persons with ID, although this kind of information was also produced, such as how many persons with ID were receiving certain benefits or housing services. For the purposes of service planning, this information is not detailed enough. As was noted in the literature review, coping of individuals with ID can vary greatly, even with the same diagnosis. Better estimates of coping might be attained if the information in these general registers was coded more appropriately. Besides levels and profiles of cognitive functions, and medical aetiological diagnoses, recording of comorbidities is essential in this respect. In order to evaluate the services delivered, such services should also be registered in detail. Ad hoc linkage of several available registers with detailed information, performed at reasonable time intervals would be most useful.

A central question might be whether or not Finland needs a national database, besides register linkage studies, as, for example, in Ireland (Kelly 2015), Western Australia (Pettersson et al. 2005, Bourke et al. 2018), and Taiwan (Lai et al. 2013). These two methods complement each other in guaranteeing that services are provided to those who need them. Both of them have different ethical problems, which

should not prevent them from helping those who need support. The great changes in social and health care reform underline the importance of this question.

In register-based studies there is always the risk of missing real cases of ID. Not only persons with cognitive performance more than 2 SD below average are in danger of being marginalised. Besides persons with general borderline competence, those with more narrow cognitive problems may have increasing difficulties in our intellectually demanding world (Whitaker 2013, Greespan et al. 2011). Therefore, longitudinal population sample studies are needed. Epidemiological research is not only motivated by practical aims to feed information into service delivery. Well-planned population sample studies can reveal new possibilities for development of prevention, care, and rehabilitation (Leonard and Wen 2002, Katusic 2017).

9 ACKNOWLEDGEMENTS

The work for this thesis was carried out at the Children's Hospital, Helsinki University Hospital and University of Helsinki.

The basis of my interest in the field of intellectual disability was laid out in the 1980's, when I worked for the whole decade in the Central Institute for the Intellectually Disabled of Helsinki. As a full-time clinician I learnt a great deal about the life of persons with ID. Personally I took care of several hundred individuals for a sufficiently long time to see many kinds of destinies. This decade left an ever-lasting influence. When dealing with stark statistical numbers I remember personalities who are behind the statistics. One of those acquaintances has lasted up to now, as at the time my family built a supporting role for a teenage boy who is now 51 years old.

I left the institution in 1990 to specialise in child psychiatry. In the latter part of that decade representatives of a newly formed association⁵ contacted the professors of child neurology, Matti Iivanainen and child psychiatry, Fredrik Almqvist. The association financed a series of surveys concerning the service systems and care for persons with ID while integration in Europe was deepening. I was appointed to the study group as the representative of child psychiatry because of my background. Among other things, we carried out some register-based surveys concerning the prevalence of ID in Finland. This started to open up the difficult field of the epidemiology of ID and we familiarised ourselves with the available registers in Finland. I am still grateful for the lengthy discussions we had at that time. The results were published by the association, and are difficult to find. The importance of that project, however, was that we had learnt how to start planning a larger register-based study after cooperation with the association was finished.

The multiple register sampling took place in 2002, concerning the year 2000. I am very grateful to the head of the statistics division of the Social Insurance Institute Kari Lindroos and the very friendly and helpful personnel for assisting me in understanding the contents of the seemingly simple registers and solving the many technical problems in register sampling. Likewise, I thank the personnel in THL and Väestörekisterikeskus for their help in building the data set. The second register sampling was performed in 2016 at THL, again with the kind help of their staff.

The research was performed alongside full-time clinical work in child psychiatry, and took a long time. There were also periods when this research was practically sleeping, especially during stressful times in my personal life, like during the end stages of my parents' lives. I am grateful for the patience of my supervisors, professor Matti Iivanainen and Dr. Markus Kaski for meetings to keep the project alive. They helpfully communicated their expertise and wisdom during these meetings. A sad break later occurred in September 2016, when Matti Iivanainen unexpectedly departed this life. Professor Eeva Aronen was then nominated as the second supervisor, just before finishing this research project.

I thank my co-authors, Professor Emeritus Fredrik Almqvist, Senior Medical Researcher Lauri J. Virta, M.D., Ph.D., and biostatistician Hannu Kautiainen for their extremely helpful cooperation.

⁵ LANEPSY ry (in Finnish, Lastenneurologisten ja psykiatristen sairauksien yhdistys), the Association for Diseases of Child Neurology and Child Psychiatry. The association did not last long, although it had a clear role.

I warmly thank the reviewers of this thesis, Professor Maria Arvio and Professor Mika Gissler for their helpful comments in clarifying the text, and Nick Bolton, Ph.D., for editing the language of the thesis.

I thank my friends and colleagues for support and encouragement. Most of all, I feel the warm love of my wife, children, grandchildren, and extended family, who have given me the motivation to combine this interesting research with very different but inspiring clinical work.

Vantaa, November 2018

Hannu

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11 APPENDIX

List of inclusion diagnoses in Studies I–III

An asterisk (*) denotes any digit. The diagnoses in brackets were accepted for inclusion only as the main diagnoses for disability pension.

ICD-9

[243*]	Congenital hypothyroidism
2775	Mucopolysaccharidosis
317*	Mild mental retardation
3180	Moderate mental retardation
3181	Severe mental retardation
3182	Profound mental retardation
319*	Unspecified mental retardation
330*	Cerebral degenerations usually manifest in childhood
7420–7422	Other congenital anomalies of the nervous system
[7423]	Congenital hydrocephalus
7580–7585	Chromosomal anomalies
[7595]	Tuberous sclerosis
[7607]	Foetal alcohol syndrome

ICD-10

E72.0	Disorders of amino-acid transport
E72.2	Disorders of sulfur-bearing amino acid metabolism
E72.5	Disorders of glycine metabolism
E75.*	Disorders of sphingolipid metabolism and other lipid-storage disorders
E76.*	Disorders of glycosaminoglycan metabolism
E77.*	Disorders of glycoprotein metabolism
E79.1	Lesch-Nyhan syndrome
E83.0	Disorders of copper metabolism
E88.8	Other specified metabolic disorders
F70.*	Mild mental retardation
F71.*	Moderate mental retardation
F72.*	Severe mental retardation
F73.*	Profound mental retardation
F78.*	Other mental retardation
F79.*	Unspecified mental retardation
F84.2	Rett's syndrome
F84.3	Other childhood disintegrative disorder
F84.4	Overactive disorder associated with mental retardation and stereotyped movements
G11.13	Early onset cerebellar ataxia (Marinesco-Sjögren)
G11.3	Cerebellar ataxia with defective DNA repair
Q00.*	Anencephaly and similar malformations
Q01.*	Encephalocele
[Q02.*]	Microcephaly
Q03.81	Hydroletharus syndrome
[Q03.*]	Congenital hydrocephalus

[Q04], [Q04.0], Q04.00, [Q04.08], Q04.2, [Q04.3], Q04.51, [Q04.8], [Q04.9]	Other congenital malformations of the brain
Q61.90	Meckel-Gruber syndrome
[Q85.1]	Tuberous sclerosis
Q86.0	Foetal alcohol syndrome (dysmorphic)
Q86.1	Foetal hydantoin syndrome
Q87.01, -.05, -.12, -.15, -.18, -.19, -.23, -.30, -.31, -.38, -.82-6	Chromosomal microdeletions, specified syndromes
Q90.*	Down's syndrome
Q91.*	Edwards' syndrome and Patau's syndrome
Q92.*, except [Q92.6]	Other trisomies and partial trisomies of the autosomes, not elsewhere classified
Q93.*	Monosomies and deletions from the autosomes, not elsewhere classified
Q97.1	Female with more than three X chromosomes
Q99.2	Fragile X syndrome
[Q99.8]	Other specified chromosome abnormalities

List of inclusion diagnoses in Study IV

An asterisk (*) denotes any digit.

E72.5	Disorders of glycine metabolism
E75.0,1,2,4,6	Disorders of sphingolipid metabolism and other lipid storage disorders
E76.0,1,2,3,8,9	Disorders of glycosaminoglycan metabolism
E77.0,1,8,9	Disorders of glycoprotein metabolism
E79.1	Lesch-Nyhan syndrome
E88.8	Other specified metabolic disorders
F70.*	Mild mental retardation
F71.*	Moderate mental retardation
F72.*	Severe mental retardation
F73.*	Profound mental retardation
F78.*	Other mental retardation
F79.*	Unspecified mental retardation
F84.2	Rett's syndrome
Q00.0	Anencephaly and similar malformations
Q03.81	Hydroletharus syndrome
Q04.00, Q04.2, Q04.51	Other congenital malformations of the brain
Q86.0	Foetal alcohol syndrome (dysmorphic)
Q87.01,05,12,15,18,19,23,30,31,38,83-6	Chromosomal microdeletions, specified syndromes
Q90.*	Down's syndrome
Q91.*	Edwards' syndrome and Patau's syndrome
Q92.*, except Q92.6	Other trisomies and partial trisomies of the autosomes, not elsewhere classified
Q93.*	Monosomies and deletions from the autosomes, not elsewhere classified
Q99.2	Fragile X syndrome

12 ORIGINAL PUBLICATIONS

Prevalence of intellectual disability: a comprehensive study based on national registers

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Abstract

Background Based on standard social benefit registers, the prevalence of intellectual disability (ID) in Finland is estimated to be 0.6%, while epidemiological surveys yield 1.1%. Combining several registers, our aim was to find a more reliable estimate of the prevalence of ID, especially among children and adolescents. This is important when special or inclusive general services are planned to meet the various needs of people with ID.

Method A survey based on eight national health and social benefit registers.

Results Combining different registers yielded a mean ID prevalence of 0.70% (95% CI 0.69–0.70%), with marked differences according to sex and age group (range 0.38–0.96%). Capture–recapture analysis gave higher prevalence estimates (range 0.57–1.08%).

Conclusions When several health and social benefit registers are surveyed, the estimated prevalence of ID increases, approaching that obtained in epidemiological surveys.

Keywords epidemiology, intellectual disability, prevalence, registers, services

Introduction

Knowing the prevalence of intellectual disability (ID) is necessary to successfully plan services for the intellectually disabled. People with ID often cannot express their needs for health and rehabilitation or other services. Thus, the supply of services, rather than being based on demand, should meet the estimated need (Taanila *et al.* 2005).

A register of all persons with ID in a certain area, with detailed information from many different perspectives, would help in estimating the need for services. There are, however, obstacles in keeping such a register. In many countries, including Finland, a conscious and determined mainstreaming policy is underway, and maintaining a register of ID people is viewed as a step backward. These registers may also be regarded as unethical and feared as a potential means of discrimination. Moreover, our experience has shown that keeping such a register up to date is demanding and the reliability is difficult to guarantee in normal clinical practice.

When social and healthcare benefits are provided to persons with ID, these are recorded in several registers. Thus, ID people may be identified from these registers according to either medical diagnosis (level or aetiology of ID) or the services supplied. However, how much standard registers help in

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estimating the actual needs of persons with ID for services remains unclear.

When a special law to guarantee rehabilitation services for persons with ID in Finland came into force in 1978, a register was founded for those being served. At intervals of a couple of years, data were updated via questionnaires sent to the service units. The last figures are from 1986, after which the collection of information was ceased. At that time, about 19 000 clients of the services (0.4% of the population) were registered (Aromaa *et al.* 1999).

The prevalence of ID has also been estimated by using registers of the National Pension Institute, which supports children with long-term illnesses and disabilities and may also grant invalidity pensions to working-aged individuals with ID. Data from this source indicate an ID prevalence of 0.3% in the age group 0–15 years and 0.6% in the age group 16–64 years.

When trying to identify persons with ID from registers, the figures acquired are normally an underestimation of the true prevalence. A cohort study, a comprehensive survey of a particular age group in a defined area, gives the most reliable estimate, but such studies are extremely labourious.

A few cohort studies with strict methodology have been undertaken in Finland. In Helsinki, the 1955 birth cohort was studied longitudinally in 1969 in a prospective manner, although retrospectively (Amnell 1974). Cases were identified from hospital birth records and they were followed through all available healthcare and social welfare records, not just registers. At 14 years of age, the cumulative incidence of ID was 1.01% and the point prevalence 0.92% (IQ range 0–67) (95% CI 0.69–1.12%), ascertained by psychological tests in every case.

In Kuopio, central Finland, the KEVA project (1978–1980) showed a prevalence of 1.38% (95% CI 1.18–1.58%) at the age of 8–9 years in a cross-sectional cohort study (Kääriäinen *et al.* 1985; Kääriäinen 1987).

In Northern Finland, two prospective birth cohort studies with a similar methodology were carried out 20 years apart (1966 and 1985–1986) (Rantakallio & von Wendt 1986; von Wendt & Rantakallio 1987; Heikura *et al.* 2003). In both cohorts, the prevalence of ID was 1.1% (95% CI 0.91–1.20% and 0.99–1.34%) at the age of 11.5 years, and in the earlier cohort 1.19% (95% CI not given) at the age

of 14 years. There was a tendency towards an increase in mild ID during this 20-year period, while severe and profound ID remained unchanged.

In an extensive review of the prevalence of ID in young people, which covered publications from 1981 to 1995, the authors came to the following conclusions (Roeleveld *et al.* 1997):

- The prevalence of severe ID is about 0.4%.
- The combined prevalence of severe and mild ID is up to 3%.
- However, due to several methodological problems, the true prevalence is difficult to state.

The POMONA project summed up epidemiological research on the prevalence of ID in the member states of the European Union (POMONA 2004). Prevalence figures depend heavily on the practical definition of ID (the role of adaptive disabilities besides measured ID in a narrow sense), and on the methodology. The POMONA report suggests that the prevalence of ID known to services is about 0.25%, while the actual prevalence of people with intellectual and adaptive disabilities may be about 1% (Whitaker 2004).

Over the last two decades, Finnish governmental reports and interest groups have assumed a prevalence of ID in Finland of 0.6% (Sosiaalialhallitus 1986; Kehitysvammaliitto 2005). Simple register-based surveys from the National Pension Institute have supported this standpoint, reporting a prevalence of 0.49% (Statistical Yearbook of Pensioners in Finland 2005). These figures are quite different from those of Finnish cohort studies and the above-mentioned reviews, casting doubt on whether these estimates are sufficiently accurate to serve as the basis for planning of services.

Our aim was to refine the estimation of the prevalence of ID and to examine whether combining several registers and using a more sophisticated analysis of data could diminish the gap between cohort studies and simple register-based surveys.

The practical aim was to determine a reliable basis for developing services for the intellectually disabled.

Methods

Data were combined from eight national registers based on unique individual social security codes

(SSC). Every person in Finland has an SSC, which is used in every register to identify individuals. We generated a list of ID diagnoses (according to both ICD-9 and ICD-10) to be used as inclusion criteria (Appendix 1). Of the aetiological diagnoses, only those in which ID is typically present were included.

A. Benefit registers of the National Pension Institute:

A1 Child Disability Allowance (CDA). The National Pension Institute pays CDA to support the care of a disabled or chronically ill child at home and to compensate for additional expenses and special arrangements. The allowance is paid regardless of the parents' or the child's income or assets.

A2 Disability Pension (DP). The DP, granted to individuals aged 16–64 years, provides compensation for working incapacity, i.e. a person's inability to engage in gainful employment, and is not intended to compensate for the actual illness or disability.

A3 Disability Allowance (DA). The DA makes it easier for disabled persons of working age (16–64 years) to manage their ordinary everyday activities and to cope with their work and studies. It is granted to persons with an illness or injury that reduces their functional capacity for a period of at least 12 months. The allowance covers costs of a general handicap, the need for assistance, services, guidance or supervision, and additional expenses.

A4 Pensioners' Care Allowance (PCA). The PCA enables pension recipients with an illness or disability to live at home, supporting their home care and reimbursing recipients for extra costs incurred by illness or disability. Persons eligible for PCA are Finnish residents who are

- aged 65 years or more; or
- aged less than 65 years but receiving a full DP, rehabilitation subsidy or individual early retirement pension.

A5 Funding of Rehabilitation (REH). The National Pension Institute grants funding for rehabilitation in conjunction with funding from other organizations. The special focus is the rehabilitation of severely handicapped people.

A6 Preferential Refunding of Long-term Medication (MED). Patients suffering from certain serious and long-term diseases are entitled to a full (100%) refund of medication expenses ('preferential refund'). One of the accepted indications is 'pertur-

bation of mind in connection with intellectual disability' (indication no. 113, case-finding criterion in this study), which was separated from 'serious psychotic and other mental disorders' (indication no. 112) in the 1960s. A precise psychiatric diagnosis is needed for indication no. 112, but not for indication no. 113, where the emphasis is on reliable verification of ID underlying the psychiatric problems. The accepted pharmaceutical preparations in this indication include antipsychotics, antidepressants and preparations for bipolar disorder and behaviour disorders.

In all but the last register, one to three diagnoses are coded for each benefit decision (according to either ICD-9 or ICD-10). The last register is based on illness-specific criteria. We identified persons who had received a refund for medication for 'perturbation of mind in connection with ID'. No diagnoses were coded in this register.

Data were collected from the year 2000. Most benefits from the National Pension Institute are long-term. Because of this and in order to confine the costs of register searching, we limited sampling in the National Pension Institute registers to 1 year.

B. Registers of Hospital Care and Care for the Intellectually Disabled, which are kept by the National Research and Development Centre for Welfare and Health (STAKES):

B1 Discharge Register of Hospitals (HOSP). All persons with an ID diagnosis who had been in hospital during 1996–2000 and were still alive in 2000 were identified. For each event, one to six diagnoses are coded in this register (according to ICD-10). As a hospital stay may be a solitary event in a person's life, a 1-year search period may be too short to identify most cases. We therefore used a period of 5 years; because the register in its current form was dated from 1996 onwards, we could not go further backwards.

B2 Discharge Register of Social Care (CARE). All persons living either permanently or temporarily during 2000 in institutions or housing units for persons with ID were identified from CARE. No diagnoses are coded in this register, but all persons have been diagnosed as intellectually disabled. Register information is gathered in two ways: upon discharge throughout the year (temporary visitors) and as a census of all inhabitants at the end of the year (temporary visitors at that time or permanent

inhabitants). Both information-gathering methods are used in institutions and in housing with 24-h assistance, but only annual census is used in less supported or independent accommodation for persons with ID.

In this study, benefits that are long-lasting and granted before the introduction of ICD-10 may have an ICD-9 diagnosis. In some cases, reapplication for the same benefit may have changed the diagnosis to an ICD-10 diagnosis. This holds true for registers A1, A2, A3, A4 and A5. For register B1, all diagnoses were derived from ICD-10.

After a combined list of all cases was generated from the separate registers, all necessary information about benefits for every index case was gathered individually from each register. The investigator was blind to the cases' identities.

The diagnosis of ID was not individually ascertained for the study population. Diagnoses were set in normal clinical practice. The total population data were received from the National Pension Institute. As a method of data ascertainment, a capture-recapture analysis was carried out based on a simple model (Tilling 2001). Confidence intervals were calculated for the proportions according to Gardner & Altman (1989).

The study was approved by the Ethics Committee of the National Research and Development Centre for Welfare and Health (STAKES).

Results

A total of 36 053 persons, 19 396 males (53.8%) and 16 657 females (46.2%), were identified as having a medical diagnosis connected with ID or another indication of ID (A6 or B2) in at least one register. The mean prevalence of ID for a total population of 5 184 980 was thus 0.70% (95% CI 0.69–0.70%); 0.77% (95% CI 0.76–0.78%) for males and 0.63% (95% CI 0.62–0.64%) for females.

Large differences in ID prevalence were present between the four age groups (≤ 15 , 16–39, 40–64, ≥ 65 years), the highest figure being 0.92% (95% CI 0.90–0.93%) in the age group 40–64 years (Table 1). In the three youngest age groups, the prevalence was higher in males than in females, but in the oldest age group the prevalence was equal.

Altogether 15 897 cases (44%) appeared in only one register (Table 2). Had only the two most important registers of the National Pension Institute (A1: CDA and A2: DP) been used, as is performed in standard prevalence estimations in Finland, only 60% of the cases would have been identified, resulting in a total prevalence 0.42% instead of 0.70%. The incremental value of adding more registers to the case finding and using a broader list of inclusion diagnoses is presented in Table 3. The relative value of different registers in case finding in each age group is also illustrated in Venn diagrams (Fig. 1).

From the Preferential Refunding of Long-term Medication (A6: MED) register, we located 2315 cases that did not appear in any other register with an ID diagnosis (Table 2). However, 999 of these appeared in one or more of the other registers of the National Pension Institute (A1: CDA, A2: DP, A3: DA, A4: PCA or A5: REH) with some other diagnoses of disability or long-term illness.

From all sources, we identified 5346 children (0–15 years) with ID, yielding an average prevalence rate of 0.53% (95% CI 0.52–0.55%). By using only the most frequently used register of A1: CDA, only 3238 children (61%) would have been found, yielding a prevalence rate of 0.32% (95% CI 0.31–0.33%).

We performed a capture-recapture analysis in which registers A1–A6 represented the capture and registers B1–B2 the recapture. This analysis increased the estimated prevalence most in the youngest and oldest age groups, by 25% and 144%, respectively, and less in the middle groups, by 12–13%. The total prevalence estimate would now be 0.87% (95% CI 0.86–0.88%), with large differences persisting between the sexes and age groups (Table 1).

Discussion

Evaluation of the results

Compared with the official administrative prevalence rate (0.6%), our estimate is higher, especially for young adults and the middle-aged. Our figure is also an administrative prevalence because all cases had been receiving benefits or care based on ID, or they had been hospitalized with an ID diagnosis.

Table 1 Prevalence of ID calculated from combined registers and capture-recapture analysis

Age (years)	Sex	Total population	Combined registers			Capture-recapture			Figures to C/R formula				
			Cases	Prevalence estimate	95% CI	Cases	Prevalence estimate	95% CI	Group A only	Group B only	Intersect A&B	New cases	Addition
0-15	M	510 852	3 121	0.61%	0.59-0.63%	3 895	0.76%	0.75-0.79%	995	930	1 196	774	25%
	F	489 831	2 225	0.45%	0.44-0.47%	2 782	0.57%	0.55-0.59%	744	634	847	557	25%
16-39	T	1 000 683	5 346	0.53%	0.52-0.55%	6 677	0.67%	0.65-0.68%	1 739	1 564	2 043	1 331	25%
	M	831 083	6 528	0.79%	0.77-0.80%	7 319	0.88%	0.86-0.90%	3 666	508	2 354	791	12%
	F	796 423	4 889	0.61%	0.60-0.63%	5 530	0.69%	0.68-0.71%	2 681	426	1 782	641	13%
	T	1 627 506	11 417	0.70%	0.69-0.71%	12 850	0.79%	0.78-0.80%	6 347	934	4 136	1 433	13%
40-64	M	889 638	8 585	0.96%	0.94-0.99%	9 614	1.08%	1.06-1.10%	5 390	512	2 683	1 029	12%
	F	888 680	7 710	0.87%	0.85-0.89%	8 669	0.98%	0.96-1.00%	4 958	446	2 306	959	12%
	T	1 778 318	16 295	0.92%	0.90-0.93%	18 282	1.03%	1.01-1.04%	10 348	958	4 989	1 987	12%
65+	M	300 174	1 162	0.39%	0.36-0.41%	2 501	0.83%	0.80-0.87%	579	407	176	1 339	115%
	F	478 299	1 833	0.38%	0.37-0.40%	4 897	1.02%	1.00-1.05%	1 112	529	192	3 064	167%
	T	778 473	2 995	0.38%	0.37-0.40%	7 296	0.94%	0.92-0.96%	1 691	936	368	4 301	144%
Total	M	2 531 747	19 396	0.77%	0.76-0.78%	23 328	0.92%	0.91-0.93%	10 630	2 357	6 409	3 932	20%
	F	2 653 233	16 657	0.63%	0.62-0.64%	21 878	0.82%	0.81-0.84%	9 495	2 035	5 127	5 221	31%
	T	5 184 980	36 053	0.70%	0.69-0.70%	45 106	0.87%	0.86-0.88%	20 125	4 392	11 536	9 053	25%

The number of missing cases (column 'new cases') in the capture-recapture analysis are calculated from the formula by Tilling (2001), which applied to the first row gives $995^{*}930/1196 = 774$. Group A refers to persons with ID who were found only in registers A1-A6 and not in registers B1-B2. Group B means the opposite. Intersect A&B refers to persons with ID who appeared in both register groups.

ID, intellectual disability.

Table 2 Numbers of cases of ID tracked from different registers (in bold) and numbers of cases occurring in only one register (in italics)

Source	0–15 years	16–39 years	40–64 years	65+ years	Total
A1: CDA	3238 <i>935</i>	263 <i>59</i>	0 <i>0</i>	0 <i>0</i>	3 501 <i>994</i>
A2: DP	0 <i>0</i>	9 117 <i>2 153</i>	13 613 <i>5 029</i>	330 <i>164</i>	23 060 <i>7346</i>
A3: DA	0 <i>0</i>	343 <i>101</i>	17 <i>12</i>	0 <i>0</i>	360 <i>113</i>
A4: PCA	0 <i>0</i>	5 933 <i>232</i>	5 803 <i>281</i>	173 <i>45</i>	11 909 <i>558</i>
A5: REH	1921 <i>207</i>	1 316 <i>146</i>	328 <i>37</i>	1 <i>0</i>	3 566 <i>390</i>
A6: MED	122 <i>4</i>	1 857 <i>84</i>	5 347 <i>863</i>	1767 <i>1364</i>	9 093 <i>2 315</i>
B1: HOSP	2770 <i>1031</i>	1 636 <i>480</i>	1 784 <i>644</i>	823 <i>643</i>	7 013 <i>2 798</i>
B2: CARE	1325 <i>410</i>	3 904 <i>410</i>	4 571 <i>291</i>	529 <i>272</i>	10 329 <i>1 383</i>
Total	5346 <i>2587 (48%)</i>	11 417 <i>3 665 (32%)</i>	16 295 <i>7 157 (44%)</i>	2995 <i>2488 (83%)</i>	36 053 <i>15 897 (44%)</i>

The totals in the bottom row are column sums for non-overlapping figures (in italics); the gross sums (in bold) are from primary data. CARE, Institutional Care for ID People; CDA, Child Disability Allowance; DA, Disability Allowance; DP, Disability Pension; HOSP, Discharge Register from Hospitals; MED, Preferential Refunding of Long-term Medication; PCA, Pensioners' Care Allowance; REH, Funding of Rehabilitation; ID, intellectual disability.

Source	Age (years) (%)				Total
	0–15	16–39	40–64	65+	
A1-A2: primary diagnoses, list 1	0.17	0.41	0.60	0.03	0.37
A1-A2: primary diagnoses, list 2	0.25	0.47	0.64	0.03	0.42
A1-A5: primary diagnoses, full list	0.32	0.57	0.69	0.04	0.48
A1-A5: all diagnoses, full list	0.37	0.63	0.81	0.05	0.56
A6: MED added	0.38	0.64	0.86	0.26	0.61
B1: HOSP added, full list	0.49	0.68	0.90	0.35	0.67
B2: CARE added-final prevalences	0.53	0.70	0.92	0.38	0.70

Table 3 Calculated prevalences of ID by age group according to the number of inclusion criteria used

A1, Child Disability Allowance; A2, Disability Pension; A3, Pensioners' Care Allowance; A4, Disability Allowance; A5, Funding of Rehabilitation; A6: MED, Preferential Refunding of Long-term Medication; B1: HOSP, Discharge Register from Hospitals; B2: CARE, Institutional Care for ID People; ID, intellectual disability.
List 1 comprises only diagnoses 317x–319x (ICD-9) and F70x–F79x (ICD-10).
List 2 comprises list 1 plus Down's syndrome added: diagnoses 758o (ICD-9) and Q90x (ICD-10).
The full list of inclusion diagnoses is presented in Appendix 1.

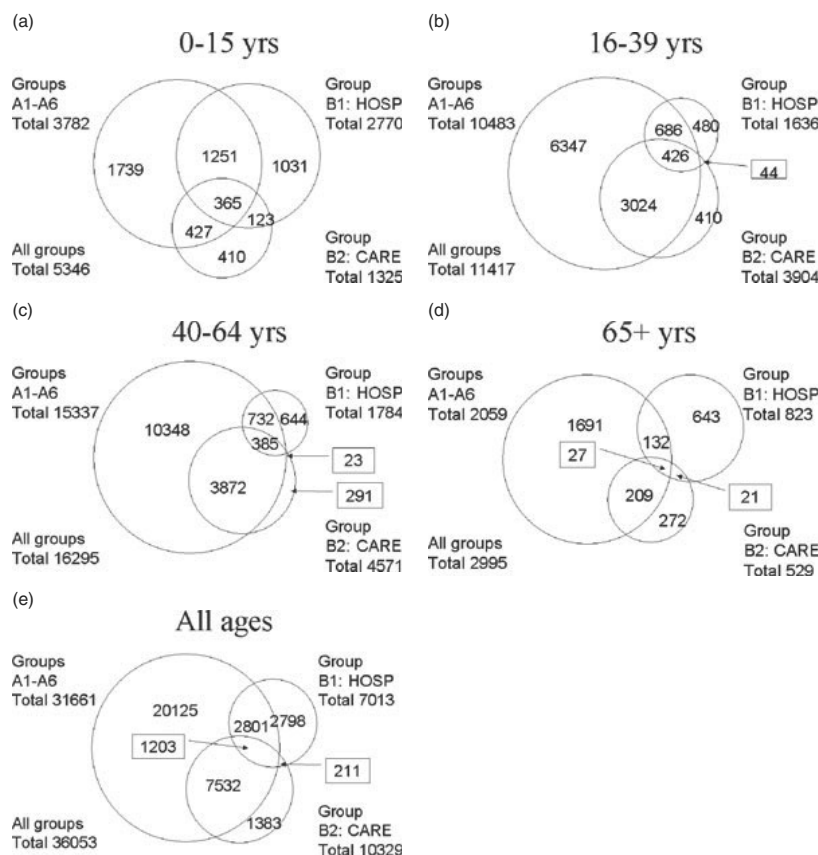


Figure 1 (a-e) Venn diagrams presenting the significance of different sources of data in finding ID persons. ID, intellectual disability; HOSP, Discharge Register of Hospitals; CARE, Discharge Register of Social Care.

Our figure represents individuals with problems in adaptation (need for a benefit) besides having a subnormal IQ (the medical criterion to receive the benefit).

The official administrative prevalence figure is based on the main benefits of the National Pension Institute (A1: CDA for children and A2: DP for adults). We determined that individuals receiving these benefits may have some other diagnosis of disability or long-term illness while simultaneously being intellectually disabled. These people are not included in the official prevalence figure. The other diagnoses will be studied in detail in a subsequent

report, but we suspect that the common concomitant handicaps of ID (epilepsy, cerebral palsy, impairments of vision and speech) will be represented. While ID is probably recognized at an individual level in these cases, when rehabilitation is arranged, it would be useful to also determine it at the official administrative level in order to develop service plans.

Compared with Finnish cohort studies, the figure acquired here for the age group 0-15 years was clearly lower and the confidence intervals did not overlap. However, the methods are not truly comparable. Whereas the cohort studies calculated

a point prevalence at a certain age (e.g. 8–9 or 14 years), we used the average administrative prevalence of all age groups between 0 and 15 years of age. If ID has been diagnosed and brought into the registers gradually over time, as we assume, then a more comparable figure might be acquired by examining the age profile year by year in this material.

The marked differences in prevalence between age groups may originate from changes in the incidence and mortality of ID, diagnostic practices and benefit provision – all of these having changed during the time period explored here. Analysing age-specific prevalences warrants further research.

Evaluation of the methods – list of inclusion diagnoses

The list of inclusion diagnoses did not contain aetiological diagnoses in which ID is not regularly present, e.g. birth complications, central nervous system infections or infantile autism. In such situations, individuals were not identified from the registers if they did not have another definitive diagnosis of ID or did not belong to the A6: MED or B2: CARE group. When the ICD-9 diagnoses were in use, the National Pension Institute often used only three digits of the diagnostic code. The diagnoses may thus remain unspecific concerning the presence or absence of ID, and we may have lost true cases by omitting those three-digit diagnoses (e.g. 758, which includes trisomies with ID and sex chromosome anomalies without ID).

Selection, reliability and coverage of the registers

The six registers of the National Pension Institute, all of which give a diagnosis of ID, were included. These registers and the two Institutional Care registers are the most commonly used for this kind of research in Finland.

There are some other potentially useful registers in Finland. The Birth Defects Register, kept by the National Research and Development Centre for Welfare and Health (STAKES), contains information about congenital anomalies and chromosomal aberrations that may be associated with ID. However, we suspected that few unique cases would be traced from this register.

Besides the national pension system, insurance companies maintain an employment pension system with separate registers. DPs are paid under both the national pension and the employment pension systems. These pension systems differ slightly with regard to the definition of disability applied. Based on our experience, we assume that practically all persons with ID receiving DP under the employment pension system also receive DP from the National Pension Institute.

The reliability of the registers utilized is a vital question. In the absence of any study evaluating their reliability, we can only present the regular updating of these registers. For the National Pension Institute to grant a benefit, proper examinations and a thorough diagnostic evaluation are necessary. However, when benefits (e.g. DP) were granted almost three decades ago, the diagnostic procedures were not the same as those today, and thus, differential diagnostic inconsistencies may exist, e.g. between psychiatric disorders and ID. We anticipate that for the younger age groups the current diagnostic evaluations are accurate, at least from the perspective of few false positives for ID. There is good reason to believe that a reliable standard also exists for the Hospital Discharge Register. In Finland, special institutional care is reserved for people with ID. Thus, the information from these registers is likely to be fairly reliable.

Disability pensions are intended as compensation for work incapacity, i.e. a person's inability to engage in gainful employment, and not as compensation for the actual illness or disability. Thus, a diagnosis of ID does not necessarily lead to a DP being granted. The same is true for Child Care Allowance. Everyone eligible for them does not receive benefits from the National Pension Institute, and not all applications for benefits are approved. Thus, the coverage is incomplete for the purposes of this study.

When a person reaches 65 years of age, he or she starts receiving Old Age Pension in place of any previous National Invalidity Pension. Because Old Age Pension does not require any medical diagnosis, ID persons 65 years or older can no longer be identified from this source, if they do not also receive Pensioners' Care Allowance (A4: PCA), which necessitates a diagnosis. This seems to be the reason that many unique cases were traced from the

register of Preferential Refunding of Long-term Medication (A6: MED) in this age group (Table 2).

At present, there is no comprehensive register of care provided to persons with ID. The register (B2: CARE) used here covers only institutional care and housing. ID persons living in housing with less than 24-h assistance could not be identified if they only visited the unit during the year. Their number is, however, low compared with those living in the unit, and most of the visitors were probably identified through other sources in this study.

The Hospital Discharge Register brings newly diagnosed and follow-up cases of ID to this material as well as cases where the diagnosis of ID has been recorded alongside another health problem. The applicable time period of 5 years to find cases from the hospital discharge register may have been insufficiently long because in many cases no repeated inpatient periods are needed after the primary investigations.

The Hospital Discharge Register concerns inpatient care. No similar register covering outpatient care in hospitals or in primary health care is available. While diagnostic investigations are most often performed in hospital inpatient wards, the possibility exists of not having found all cases, as a number of outpatients are missing from the statistics.

A definite diagnosis of ID is sometimes postponed, especially in young age groups. The reasons for this are manifold: difficulty in examining the developmental level, additional handicaps (epilepsy, cerebral palsy, etc.) as the stated diagnoses and targets of rehabilitation, or unwillingness to reveal the presence of ID to the family.

We suspect that all of these facts are reflected in the result of capture–recapture analysis, which raised the prevalence estimates of ID considerably, especially in the youngest and oldest age groups. However, the necessary assumptions for a simple capture–recapture method are violated in this material (Tilling 2001). First, the two sources are interdependent in many ways, e.g. hospital investigations are used for applying for a disability benefit. Second, individuals do not have the same probability of ‘being captured’, i.e. being included in a register, because of stringent criteria. For these reasons, we hesitate to use a more complicated capture–recapture model (Hook & Regal 1997). Instead, we suppose that analysing the content of

the data in more detail, by 1-year age cohorts, geographically, by diagnostic groups, and so identifying possible reasons for missing cases may be more useful. Although some authors have suggested that a total prevalence estimate should not be derived from the pooled count of different sources, but should also estimate the number of missed cases (Laporte 1994), we consider the pooled count to be a more reliable estimate statistically.

However, for the above-mentioned reasons, the estimate of the prevalence of ID based on pooled count is more probably an underestimate than the opposite – an underestimate concerning both the ‘true prevalence’ and those in need of ID services.

This method of studying the ID population has some definite advantages: it is easy to perform and privacy rights are not violated as the individuals are not known to the researcher.

Conclusions

Since services targeted to the population-at-large are not always readily available to those with ID, a system of special care, positive discrimination, is needed. This kind of multiple register survey may help in identifying social needs by uncovering statistics from different sources. The true prevalence of people with intellectual and adaptive disabilities remains, however, elusive when only registered cases are considered.

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Appendix I

List of inclusion diagnoses used in case finding. The letter 'x' after the code represents any alternative. Those marked by an asterisk in front of the code were only used as inclusion criteria for the main diagnosis for disability pension (A2: DP).

ICD-9

*243x	Congenital hypothyroidism
2775	Mycopolysaccharidoses
317x	Mild mental retardation
3180	Moderate mental retardation
3181	Severe mental retardation
3182	Profound mental retardation
319x	Unspecified mental retardation
330x	Cerebral degenerations usually manifest in childhood
7420–7422	Other congenital anomalies of the nervous system
*7423	Congenital hydrocephalus
7580–7585	Chromosomal anomalies
*7595	Tuberous sclerosis
*7607	Foetal alcohol syndrome

ICD-10

E72.0	Disorders of amino-acid transport
E72.2	Disorders of sulphur-bearing amino acid metabolism
E72.5	Disorders of glycine metabolism
E75.x	Disorders of sphingolipid metabolism and other lipid storage disorders
E76.x	Disorders of glycosaminoglycan metabolism
E77.x	Disorders of glycoprotein metabolism
E79.1	Lesch-Nyhan syndrome
E83.0	Disorders of copper metabolism
E88.8	Other specified metabolic disorders
F70.x	Mild mental retardation
F71.x	Moderate mental retardation
F72.x	Severe mental retardation

F73.x	Profound mental retardation	Q61.90	Meckel-Gruber syndrome
F78.x	Other mental retardation	*Q85.1	Tuberous sclerosis
F79.x	Unspecified mental retardation	Q86.0	Foetal alcohol syndrome (dysmorphic)
F84.2	Rett's syndrome	Q86.1	Foetal hydantoin syndrome
F84.3	Other childhood disintegrative disorder	Q87.01, -.05, -.12, -.15, -.18, -.19, -.23, -.30, -.31, -.38, -.82-6	Chromosomal microdeletions. specified syndromes
F84.4	Overactive disorder associated with mental retardation and stereotyped movements		
G11.13	Early onset cerebellar ataxia (Marinesco-Sjögren)	Q90.x	Down's syndrome
G11.3	Cerebellar ataxia with defective DNA repair	Q91.x	Edwards' syndrome and Patau's syndrome
Q00.x	Anencephaly and similar malformations	Q92.x, except *Q92.6	Other trisomies and partial trisomies of the autosomes, not elsewhere classified
Q01.x	Encephalocele	Q93.x	Monosomies and deletions from the autosomes, not elsewhere classified
*Q02.x	Microcephaly	Q97.1	Female with more than three X chromosomes
Q03.81	Hydrolethrus syndrome	Q99.2	Fragile X syndrome
*Q03.x	Congenital hydrocephalus	*Q99.8	Other specified chromosome abnormalities
*Q04, *Q04.0, Q04.00, *Q04.08, Q04.2, *Q04.3, Q04.51, *Q04.8, *Q04.9	Other congenital malformations of the brain		

Age-specific prevalence of intellectual disability in Finland at the beginning of new millennium – multiple register method

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Abstract

Background In the national study of multiple registers in 2000, the average prevalence of intellectual disability (ID) was 0.70%, with marked differences by age group (range 0.38–0.96%) – what are these differences in detail, and can they be understood?

Method This study was based on two national health registers and six social benefit registers. Prevalence of ID was calculated by 1-year age cohorts.

Results The multiple register prevalence of ID increased steadily from 0.20% in the first life year to 0.74% (male: 0.90%, female: 0.58%) at 10 years. For boys, the rate fell to 0.71% at 11 years. For both sexes, a steady increase was noted in the distribution up to 40 years (male: 0.84%, female: 0.73%), followed by a sharper increase to the maximum prevalence (male: 1.19% at 48 years, female: 1.05% at 50 years). At the pension age of 66 years, a sudden drop to 0.49% occurred for men and women. Different registers gave very different age distributions.

Conclusions By examining the data by 1-year age cohorts, and by understanding the role of each reg-

ister, it could be deduced that a proportion of cases in younger age groups is lacking, and a remarkable proportion of elderly ID persons is missing from the pooled data. The findings were more difficult to interpret, if the data were grouped into bigger age groups.

Keywords intellectual disability, methodology in research, prevalence, use of registers

Introduction

In our multiple register study of intellectual disability (ID) (Westerinen *et al.* 2007), the average prevalence of ID was 0.70% (95% CI 0.69% to 0.70%), with marked differences (range 0.38–0.96%) between the age groups 0–15 years, 16–39 years, 40–64 years and 65+ years. What lies behind these group differences, and can they better be understood, if the prevalence is calculated by 1-year age cohorts? In other published studies, big differences by age group have been noticed too, but without any consistent mode.

In Denmark, the prevalence of ID has been surveyed three times over 140 years (1855, 1965 and 1979 – see Dupont 1989) on the basis of nationwide registration. The data are shown for 5-year categories. In each survey, the distribution had a peak

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value (approximately 0.7%), at 10–15 years in 1855, and later around 20 years. After the peak value, the prevalence decreased rather linearly. Even in the oldest age group (90+ years), the prevalence was still approximately 0.2% in 1965 and in 1979.

In Ireland, the National Intellectual Disability Database Committee has published annual reports from the ID database during 1999–2010 (Ireland Health Research Board n.d.). This database is based on a voluntary registration to receive services for ID. No studies have been conducted on the coverage of the database, but the willingness to register for organised services is presumed to be high. In data from 2009 (Kelly *et al.* 2010), the peak prevalence (1.00%) was reached in the age group 10–14 years, but in previous years' analyses the peak prevalence was more often in the age group 15–19 years. After the peak value, the prevalence has decreased: in the 2009 data, there was another minor peak in the age group 35–54 years, but in the data from 1999 to 2006 the decrease has been continuous. In the oldest age group, 55+ years, the prevalence was approximately 0.35% throughout the study period.

In Taiwan, the prevalence of ID in different age groups was surveyed annually in 2000–2007 (Lin 2009). In the most recent data in 2007, the peak prevalence (0.69%) was reached in the age group 15–17 years. In the older age groups, the prevalence decreased first steeply to a level of 0.40% in the age group 30–44 years, and the more slowly. In the oldest age group, 65+ years, the prevalence was 0.11%.

In the Netherlands, the prevalence of ID was determined in the province of Limburg in 2001, finding cases through several sources with good coverage (van Schroyen Lantman-de Valk *et al.* 2006). The prevalence was between 0.63% and 0.85% in all four age groups between 5 and 69 years. The highest value was in the age group 35–49 years. In the oldest age group, 70+ years, it was still 0.20%. This study differed markedly from others that had a single, marked peak in adolescence, and much lower figures in middle-aged subjects.

Although 25 studies were presented on the prevalence of ID with age distribution in a recent meta-analysis (Maulik *et al.* 2011), we could not find a single study in which the age distribution was shown in 1-year age cohorts, rather than in broader

age groups. Moreover, most of these studies contained subgroups of ID persons, for example severe ID, or only children, and not the whole ID population in a certain area. None of the studies did examine whether there are inconsistencies in the defined age distribution.

Theoretical assumptions about the age-specific prevalence distribution of intellectual disability

From a purely theoretical viewpoint, we can construct what an age-specific prevalence distribution would look like if it is based on a set of defined premises. We consider the following set of premises:

- 1 On the basis of the definition of ID, the condition should be diagnosed during developmental age, that is, before the age of 18 years, until which age the prevalence should reach its peak level.
- 2 Intellectual disability can be diagnosed immediately after birth only in a minority of cases (e.g. Down's syndrome and other chromosomal or congenital conditions). Most diagnoses are, however, made later in the early years.
- 3 Mortality is higher among persons with ID than in the general population. The effect of higher mortality begins to have an influence on the age-specific prevalence of ID first slowly, but then more markedly in the middle ages, especially in severe ID, and in Down's syndrome where Alzheimer's dementia shortens life expectancy remarkably.

If the prevalence among young adults is 1%, these premises would produce a distribution similar to that in Fig. 1. If the prevalence is lower, the form of the distribution is nevertheless the same. Any differences in observed distribution that differ markedly from those expected warrant an explanation.

As potential explanatory factors for observed differences in this kind of multiple register study, we can discuss temporal changes in the following:

- incidence of ID;
- recognition of developmental delay in health care resulting in commencement of clinical investigations;
- diagnostic practices of ID;
- arrangement of services for ID persons with case registration; and
- survival of persons with ID compared with the general population (due to diseases, wars, etc.).

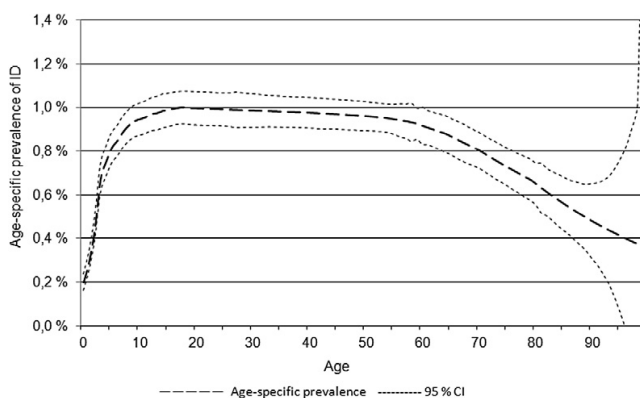


Figure 1 Theoretical age-specific prevalence distribution based on the premises described in text. The 95% confidence interval is calculated for the proportion in a population whose structure matches that of Finland in the year 2000. ID, intellectual disability.

All of these factors may change from time to time due to changes in medical practices, in the care of ID persons, or in society in general, thus having a manifold sum effect on the age distribution of ID populations.

In this study, we examine the differences in age-specific prevalence of ID, compare the observed distribution with the expected one, consider possible reasons for the discrepancy and discuss the influence of the findings on the reliability of the register-based prevalence estimate of ID.

Methods

We utilised data from eight national registers.

A. Benefit registers maintained by the Social Insurance Institution, including ID recipients:

A1. Child Disability Allowance (CDA). CDA is intended to support the care of a disabled or chronically ill child at home and to compensate for additional expenses and special arrangements. CDA is paid only to outpatients, not to those in long-term institutional care. Eligibility for CDA is not dependent on a family's socio-economic situation or place of residence.

A2. Disability Pension (DP). The DP, granted to individuals aged 16–64 years, provides compensation for working incapacity, that is, a person's inability to engage in gainful employment, and is not intended to compensate for the actual illness or disability.

A3. Disability Allowance (DA). The DA makes it easier for rather mildly disabled persons of working

age (16–64 years) to manage their ordinary everyday activities and to cope with their work and studies. It is granted to persons with an illness or injury that reduces their functional capacity for a period of at least 12 months. The allowance covers costs of a general handicap, the need for assistance, services, guidance or supervision, and additional expenses. DA is not paid to those receiving DP or those in long-term institutional care.

A4. Pensioners' Care Allowance (PCA). The PCA enables pension recipients with an illness or disability to live at home, supporting their home care and reimbursing recipients for extra costs incurred by illness or disability. Persons eligible for PCA are Finnish residents who are

- aged 65 years or more; or
- aged 16–64 years receiving a full disability pension, rehabilitation subsidy or individual early retirement pension.

A5. Funding of Rehabilitation (REH). The Social Insurance Institution grants funding for rehabilitation in conjunction with funding from other organisations. The special focus is the rehabilitation of severely handicapped persons.

A6. Preferential Refunding of Long-term Medication (MED). Patients suffering from certain serious and long-term diseases are entitled to a full (100%) refund of medication expenses ('preferential refund', PR). One of the accepted indications is 'perturbation of mind in connection with ID' (indication #113, case-finding criterion in this study). The accepted pharmaceutical preparations in this indication include antipsychotics, antidepressants

and preparations for bipolar disorder and behaviour disorders. The condition of ID has to be verified in addition to the psychiatric co-morbidity.

In all but the last register, one to three diagnoses are coded for each benefit decision (according to either ICD-9 or ICD-10).

Data were collected from the year 2000. The benefits from the Social Insurance Institution are in the majority of cases long-term. Because of this and in order to restrict the costs of register searching, we limited sampling in the Social Insurance Institution registers to 1 year.

B. Registers of Hospital Care and Care for the Intellectually Disabled, which are kept by the National Institute for Health and Welfare:

B1. Discharge Register of Hospitals (HOSP). All persons with an ID diagnosis who had been in hospital during 1996–2000 and were still alive in 2000 were identified. For each event, one to six diagnoses are coded in this register (according to ICD-10). As a hospital stay may be a solitary event in a person's life, a 1-year search period may be too short to identify most cases. We used a period of 5 years; the register in its current form was dated from 1996 onwards, and thus we could not go further backwards.

B2. Discharge Register of Social Care (CARE). All persons living either permanently or temporarily during 2000 in institutions or housing units for persons with ID were identified from CARE. No diagnoses are coded in this register, but all persons have been diagnosed as intellectually disabled. Register information is gathered in two ways: upon discharge throughout the year (temporary visitors) and as a census of all inhabitants at the end of the year (temporary visitors at that time or permanent inhabitants). Both information gathering methods are used in institutions and in housing with 24-h assistance, but only annual censuses are used in less supported or independent accommodation for persons with ID.

In this study, benefits that are long-lasting and granted before the introduction of ICD-10 may have an ICD-9 diagnosis. In some cases, reapplication for the temporarily granted benefit may have changed the diagnosis to an ICD-10 diagnosis. This holds true for registers A1, A2, A3, A4 and A5. For register B1, all diagnoses were derived from ICD-10.

Finland has a National Health Insurance Scheme funded from tax revenue; it covers all 5.18 million residents (in 2000) and is governed by the Social Insurance Institution. Each beneficiary has a unique individual social security code, including date of birth and gender, and registry linkage was based on this identifier preventing double counts. We generated a list of ID diagnoses (according to both ICD-9 and ICD-10) to be used as inclusion criteria (Appendix 1). Of the aetiological diagnoses, only those in which ID is typically present were included.

After a combined list of all cases was generated from the separate registers, all necessary information about benefits for every index case was gathered individually from each register. The investigators were blind to the cases' identities.

Diagnoses of ID had been set in normal clinical practice. The prevalence rates of ID were calculated by dividing the number of patients (retrieved from registers) by the population at risk (per 100 persons). The group at risk consisted of the whole Finnish population (5.18 million) at the end of 2000 (Official Statistics of Finland n.d.). The 95% confidence intervals (95% CI) for prevalence rates were calculated for the proportions according to Gardner & Altman (1989).

The age distribution of the whole Finnish population in 2000 is shown in Fig. 2. Marked troughs were present in the age cohorts 56–58 and 60 years. These are due to the Finnish Winter War (1939–1940) and the Continuation War (1941–1944), when birth rates were low. After these wars, by contrast, the birth rate increased for several years, resulting in the largest age cohorts ever in Finland (born in 1945–1949).

The study protocol was approved by the Ethics Committee of the National Institute for Health and Welfare.

Results

In the multiple register study, we identified 36 053 persons afflicted with ID, 19 396 male and 16 657 female. This places the nationwide prevalence rate of ID at 0.70% (95% CI 0.69% to 0.70%). The age distribution of ID persons (numbers of cases) is shown in Fig. 3 and the distribution of age-specific prevalence of ID in Fig. 4.

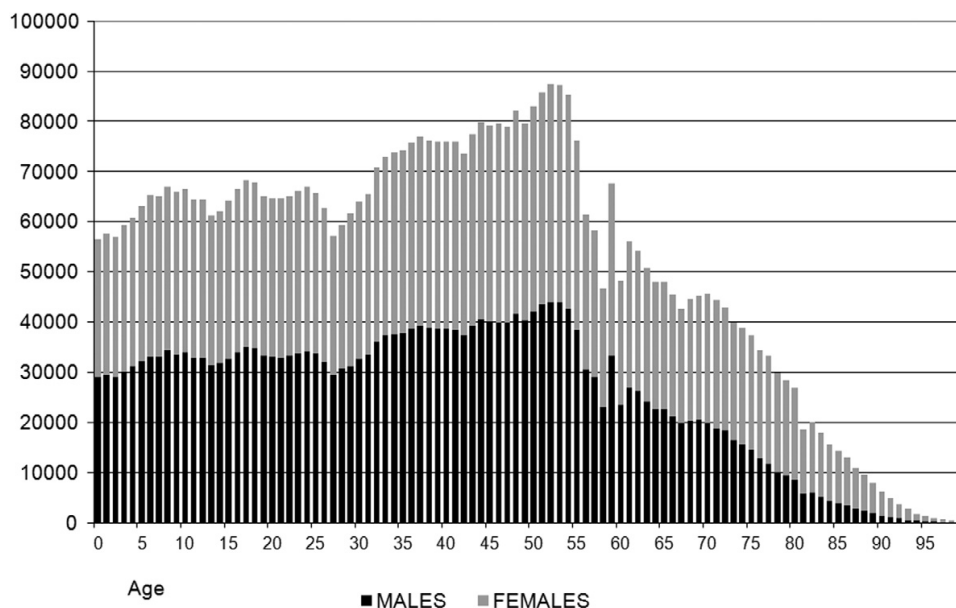


Figure 2 Age distribution of the Finnish population of 5.2 million in 2000.

The relative importance of different registers for identifying cases is shown in Fig. 5.

Age group 0–15 years

The multiple register prevalence of ID increased steadily from 0.20% (95% CI 0.16% to 0.23%) in the first life year to 0.74% (95% CI 0.68% to 0.81%) at 10 years: male 0.90% (95% CI 0.80% to 1.00%) and female 0.58% (95% CI 0.50% to 0.66%). For boys, the rate fell to 0.71% (95% CI 0.62% to 0.80%) at 11 years.

The ID cases under 16 years of age came mainly from the registers A1:CDA and B1:HOSP (Fig. 5). The registers serve very different purposes. Register A1:CDA contains all those who have received the benefit (Child Disability Allowance), often long-term, until the age of 16 years, while register B1:HOSP contains all those who have been examined in hospital either for basic diagnostic purposes or during check-up visits.

In this study, we searched the B1:HOSP register from its inception in 1996 and combined all cases from the 5-year period 1996–2000, with the requirement of the case being alive in 2000. From the 2770 ID persons found in this register, only 1575 (49.7%) had appeared in the A1:CDA register.

Age group 16–64 years

The prevalence distribution between 16 and 64 years can be divided into three different parts. Between the ages 16 and 30 years, the prevalence seemed to be rather stable (individuals born 1969–1983). Between the ages 30 and 50 years, the prevalence rose from 0.67% to 1.07% (individuals born 1950–1970). Thereafter, the distribution was U-shaped until the age of 65 years, with minimum prevalence of 0.82%, then rising again to 0.94%.

In this age group, 82.0% of the cases could be traced from the A2:DP register. The rest of the cases were traced rather evenly from the other registers relevant to this age group.

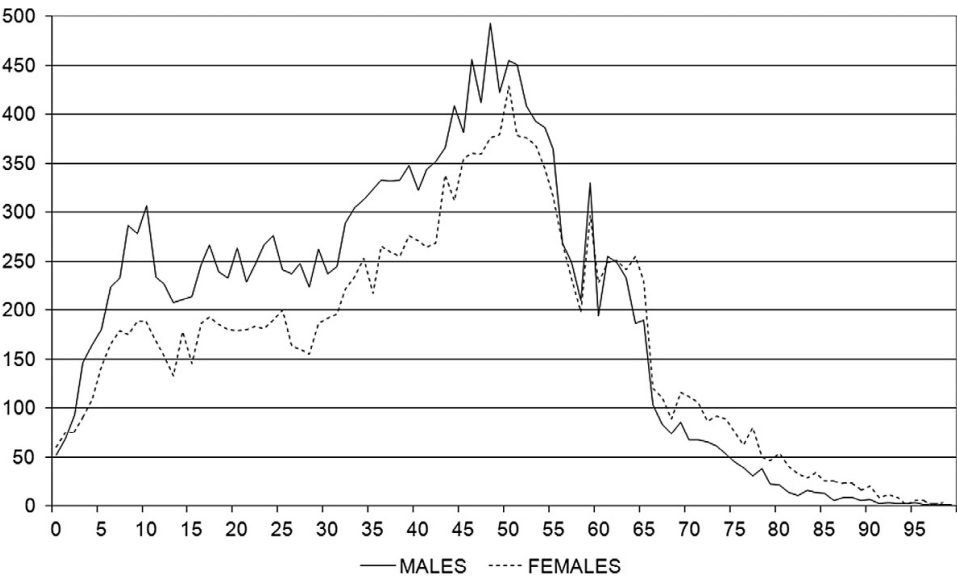


Figure 3 Age distribution of cases with intellectual disability.

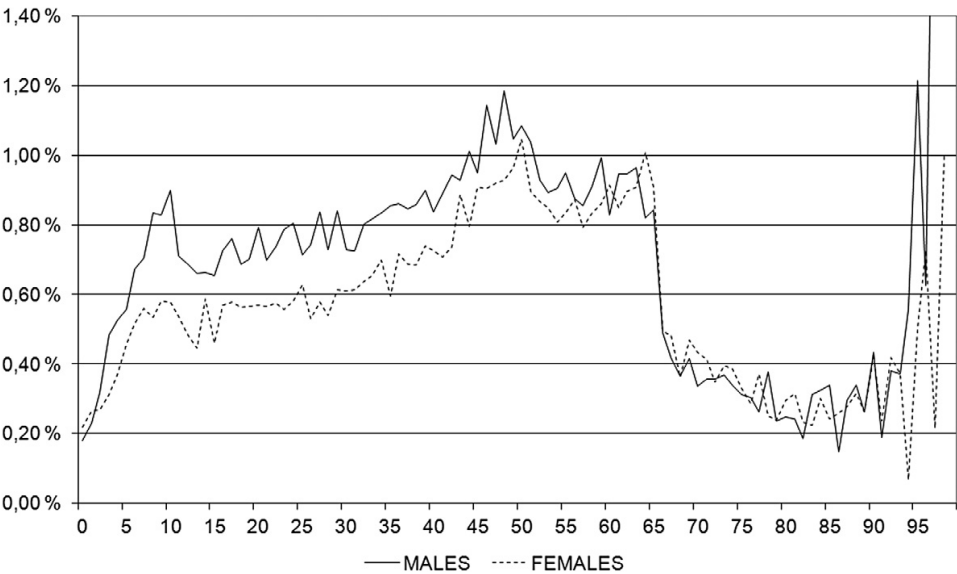


Figure 4 Distribution of age-specific prevalence in multiple register study of intellectual disability.

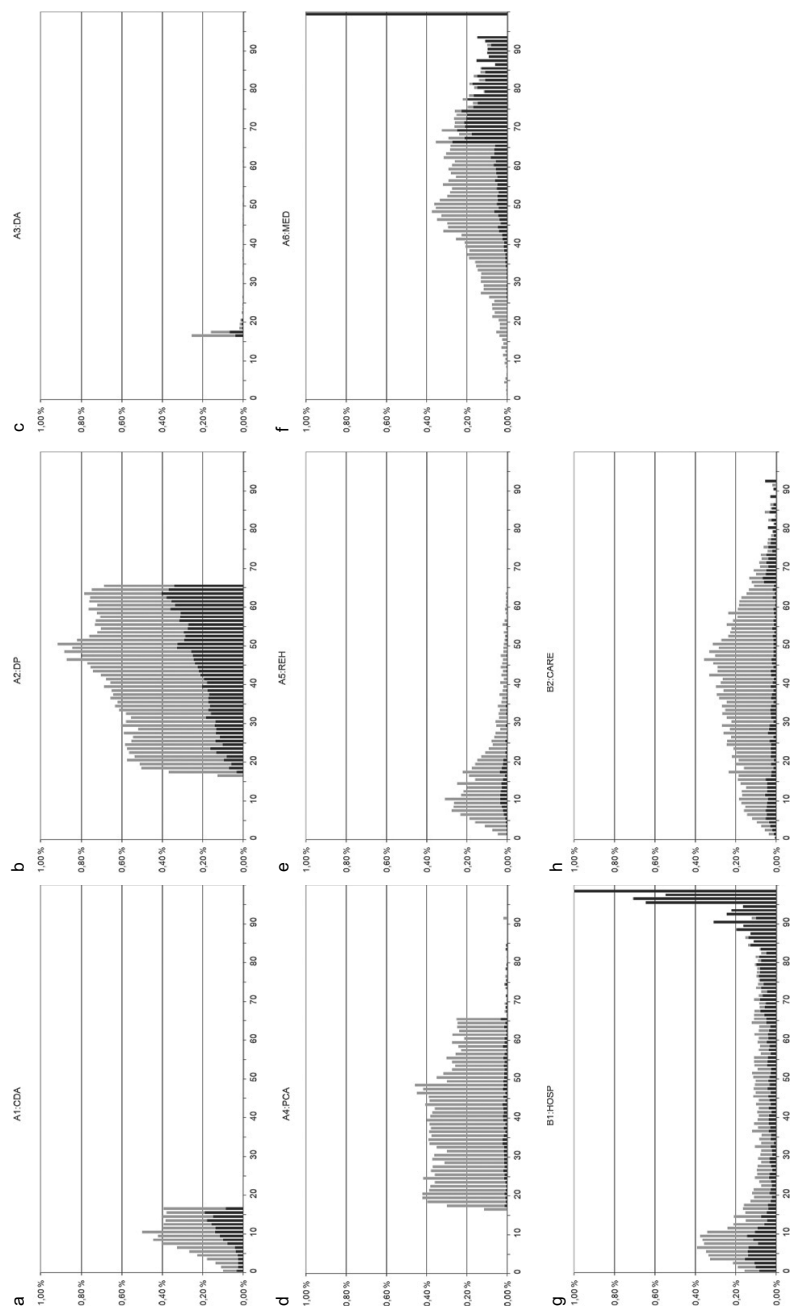


Figure 5 Age-specific prevalence of intellectual disability in different registers of the multiple register study. The black columns are based on the unique cases, and grey on total number of cases found in that register. A1:CDA, Child Disability Allowance; A2:DP, Disability Pension; A3:DA, Disability Allowance; A4:PCA, Pensioners' Care Allowance; A5:REH, Funding of Rehabilitation; A6:MED, Preferential Refunding of Long-term Medication; B1:HOSP, Discharge Register of Hospitals; B2:CARE, Discharge Register of Social Care.

Age group 65+ years

After the pension age of 65 years there was a sudden drop in prevalence to 0.49% (95% CI 0.43% to 0.55%). Until this age, the cases were mainly found from the A2:DP register, but the benefit ceases at the age of 65 years. The persons are then transferred to the old age pension, which does not depend on diagnosis, and the previous diagnoses are thus lost.

The proportion of ID persons traced uniquely via registers A6:MED and B2:CARE increased abruptly at the age of 66, at the same time as they could no more be traced via A2:DP. From this we can deduce that a sizable proportion of elderly ID persons is missing from our multiple register study.

Discussion

The investigation of the prevalence distribution by 1-year cohorts and by sex revealed several new and unexpected significant findings.

Age group 0–15 years

We do not have any studies for reference concerning the rate by which ID is diagnosed, and which is reflected in the age-specific prevalence distribution of early years. However, this seems to follow expectations, when we take into consideration the fact that many aetiologies of ID can be recognised during childhood, not soon after birth. Even before ID is recognised, many persons have reached the services on the basis of co-morbidities, like cerebral palsy or epilepsy.

After a steady increase, relative peak prevalence was noted at the age of 10 years, not at the end of the developmental age, and a sudden drop occurred after the peak prevalence in boys. The incidence might be speculated to have been exceptionally high in the cohort aged 10 years. However, our analysis of the origin of the cases in the registers does not support this hypothesis. After recognition of ID, only half of the persons start receiving long-term benefits on the basis of ID – thus, the remainder cannot be found in any of the registers.

Age group 16–64 years

For both sexes, there was first a steady but then an accelerating increase in prevalence distribution until

a maximum was reached (male: 1.19% at 48 years, female: 1.05% at 50 years). We can again present several hypotheses to explain this finding. First, the incidence may have been higher during 1948–1955. This hypothesis is interesting because the age cohorts were born after the wars, in 1945–1949, little after the so-called baby boomers (see Fig. 2). If we knew more about the changes in the aetiological diagnoses, we might better evaluate the support for this hypothesis. Our data do not, however, permit such an analysis.

Second, ID persons in this age group may appear more by age in the registers due to the need of certain benefits. This hypothesis is contradicted by the benefits (in 80% of cases A2:DP) most often being granted in early adulthood (Westerinen 2012, unpublished data).

Third, the criteria for ID may have been less strict during the time when the diagnoses were given to the bigger age cohorts. No data exist for or against this hypothesis. However, we know that decades ago many ID persons who also had severe mental disorders (high co-morbidity) were placed in mental hospitals without the ID being diagnosed. During the 1980s mental hospitals were inspected, and several ID persons were identified and diagnosed on the basis of anamnestic records from the developmental years, but in many cases the possibility of verifying the diagnosis of ID was already lost.

Age group 65+ years

An abrupt drop occurred in the prevalence of ID after 65 years. Could this be due to higher mortality? Although mortality is higher for ID persons than in the general population, this cannot explain such an abrupt drop. Instead, ID persons disappear from the registers, while the old age pension needs no diagnosis. In this oldest age group, more cases are missing than in the other age groups. Thus, there is the possibility of underestimating the elderly ID persons' needs for services, especially when the age cohorts with higher prevalence of ID get older.

The observed sex ratio, overrepresentation of men until an older age, is consistent with the literature (e.g. van Schrojenstein Lantman-de Valk *et al.* 2006).

Total population estimate of the prevalence of intellectual disability

Correcting the whole population prevalence estimate concerning the youngest and oldest age groups might yield a somewhat higher figure than previously estimated (Westerinen *et al.* 2007). The corrected figure would nevertheless be markedly lower than in Finnish cohort studies of children (Amnell 1974; Rantakallio and von Wendt 1986; Kääriäinen 1987; Heikura *et al.* 2003 – all reviewed in Westerinen *et al.* 2007) – or international estimates (Roeleveld *et al.* 1997; Whitaker 2004; Maulik *et al.* 2011). It is very likely that local cohort studies find all the cases, especially the milder ones, much more thoroughly than national register-based studies. There is also the possibility that the incidence of ID has been showing a decreasing tendency from a previous level of approximately 1% to approximately 0.7–0.8% during the decades 1940–1970, which can now be seen in the prevalence distribution by age. This holds true only if there are no major unidentified sources of underrepresentation of cases in the registers that we used. A follow-up study with the present methodology might yield further information.

Compared with earlier studies, the prevalence distribution of ID here resembled most the figures for Limburg, the Netherlands (van Schrojenstein Lantman-de Valk *et al.* 2006).

Strengths and limitations

The list of inclusion diagnoses did not contain aetiological diagnoses in which ID is not regularly present, for example, birth complications, central nervous system infections or infantile autism. In such situations, ID individuals were not identified from the registers if they did not have another definitive diagnosis of ID or did not belong to the A6:MED or B2:CARE group. This is a potential source of underestimation of the prevalence of ID.

When the ICD-9 diagnoses were in use, clinicians occasionally used only three digits of the diagnostic code. The diagnoses may thus remain unspecific concerning the presence or absence of ID, and we may have lost true cases by omitting those three-digit diagnoses (e.g. 758, which includes trisomies

with ID and sex chromosome anomalies without ID). There were 71 persons with an unspecific diagnosis 758, who were not included in the ID study group. Even if they all were ID persons, the observed prevalence estimate would have increased only insignificantly.

Although our inclusion criteria included only those autism spectrum disorders (ASD), where ID is regularly present (F84.2 – Rett's syndrome, F84.3 – Other childhood disintegrative disorder and F84.4 – Overactive disorder associated with mental retardation and stereotyped movements), we also searched all persons with any ASD diagnoses (F84.x) to check how many of them were included with a diagnosis of ID. The age distribution of persons with all ASD diagnoses was very narrow, with a maximum prevalence of 0.43% at the age of 8 years. Above 20 years the prevalence was under 0.05%. The proportion of ASD persons also having a diagnosis of ID was 25% in the age group 0–15 years. This figure compares well with that of Pinborough-Zimmerman *et al.* (2011). Thus, we may suppose not many intellectually disabled persons with ASD diagnosis, but without an actual ID diagnosis, would have been missed from our study.

Selection and coverage of the registers were discussed in our previous paper. It was noted that the estimate of the prevalence of ID based on pooled count is more probably an underestimate than overestimate. Although sampling was performed at one time point from the registers (except from B1:HOSP, where sampling was performed from five consecutive years), the fact that the benefits are normally allowed for several years lessens the probability of case loss.

Those ID persons most probably not being found to our study group are mildly disabled, who did not receive child care allowance (A1:CDA) or disability pension (A1:DP), nor the services for ID persons (B2:CARE) and did not appear in the hospital discharge register (B1:HOSP). They might receive the benefits but with non-ID diagnoses, like co-morbidity. The conclusion for the health-care services is that the diagnosis of ID should be registered in these occasions, if it is set. While the estimation of service needs for ID persons relies on this kind of statistics, it is best practice, and the interests of the ID people not to hide the diagnosis.

The present study strengthened the multiple register method by showing the relative importance of different registers in different age groups, and showing possible inconsistencies. The findings also open new important questions concerning, for example, changes in prevalence along time, which can be further studied using the same registers longitudinally.

Conclusions

The 1-year age distribution showed several unexpected relative low and high values. Understanding the role of different registers, it could be deduced that a proportion of cases in younger age groups is lacking, and a remarkable proportion of elderly ID persons is missing from the pooled data. These findings were not possible, if the data were grouped to bigger age groups.

During developmental age there seems to be a lag in recording an obvious diagnosis of ID for several reasons. Even when ID is noted in hospital examinations, it is not used as a basis for benefits: instead, diagnoses of several co-morbidities are used. The ID diagnosis should also be recorded because the needs of ID persons should be recognised at a service planning level.

The prevalence distribution by age gives a deeper understanding of the role of the different registers for identifying cases of different age groups. More than grouped data, it shows important interactions of the ID population with the national infrastructure of services and supports. It also provides information about how changes in the prevalence of ID in different age groups might be followed easier than using the multiple register study.

Acknowledgements

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Appendix I

List of inclusion diagnoses used in case finding. The letter 'x' after the code represents any alternative. Those marked by an asterisk in front of the code were only used as inclusion criteria for the main diagnosis for disability pension (A2:DP)

ICD-9

*243x	Congenital hypothyroidism
2775	Mycopolsaccharidoses
317x	Mild mental retardation
3180	Moderate mental retardation
3181	Severe mental retardation
3182	Profound mental retardation
319x	Unspecified mental retardation
330x	Cerebral degenerations usually manifest in childhood
7420–7422	Other congenital anomalies of the nervous system
*7423	Congenital hydrocephalus
7580–7585	Chromosomal anomalies
*7595	Tuberous sclerosis
*7607	Fetal alcohol syndrome

ICD-10

E72.0	Disorders of amino acid transport
E72.2	Disorders of sulphur-bearing amino acid metabolism
E72.5	Disorders of glycine metabolism
E75.x	Disorders of sphingolipid metabolism and other lipid storage disorders
E76.x	Disorders of glycosaminoglycan metabolism

E77.x	Disorders of glycoprotein metabolism
E79.1	Lesch–Nyhan syndrome
E83.0	Disorders of copper metabolism
E88.8	Other specified metabolic disorders
F70.x	Mild mental retardation
F71.x	Moderate mental retardation
F72.x	Severe mental retardation
F73.x	Profound mental retardation
F78.x	Other mental retardation
F79.x	Unspecified mental retardation
F84.2	Rett's syndrome
F84.3	Other childhood disintegrative disorder
F84.4	Overactive disorder associated with mental retardation and stereotyped movements
G11.13	Early onset cerebellar ataxia (Marinesco–Sjögren)
G11.3	Cerebellar ataxia with defective DNA repair
Q00.x	Anencephaly and similar malformations
Q01.x	Encephalocele
*Q02.x	Microcephaly
Q03.81	Hydrolethrus syndrome
*Q03.x	Congenital hydrocephalus
*Q04, *Q04.0, Q04.00, *Q04.08, Q04.2, *Q04.3, Q04.51, *Q04.8, *Q04.9	Other congenital malformations of the brain
Q61.90	Meckel–Gruber syndrome
*Q85.1	Tuberous sclerosis
Q86.0	Fetal alcohol syndrome (dysmorphic)
Q86.1	Fetal hydantoin syndrome
Q87.01, -.05, -.12, -.15, -.18, -.19, -.23, -.30, -.31, -.38, -.82-6	Chromosomal microdeletions, specified syndromes
Q90.x	Down's syndrome
Q91.x	Edwards' syndrome and Patau's syndrome
Q92.x, except *Q92.6	Other trisomies and partial trisomies of the autosomes, not elsewhere classified
Q93.x	Monosomies and deletions from the autosomes, not elsewhere classified
Q97.1	Female with more than three X chromosomes
Q99.2	Fragile X syndrome
*Q99.8	Other specified chromosome abnormalities

BRIEF REPORT

The prevalence of intellectual disability among 1-year cohorts of Finnish elderly as determined with the multiple register method

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ABSTRACT

Background Longevity is increasing among persons with intellectual disability (ID) simultaneously with that of the general population. There is a need to update prevalence figures of ID among the elderly. The aim of this study was to estimate the prevalence of ID among the elderly in Finland. **Method** We combined data from 8 national health and social benefits registers to calculate the ID prevalence for age cohorts separated by 1-year intervals. We corrected a discontinuity due to loss of diagnostic information in the main registers at retirement age (65 years) by comparing the age distribution to that of the entire population.

Results The distribution correction more than doubled the number of persons with ID in this age group, resulting in an apparent prevalence of 0.75%.

Conclusions The 1-year interval between age cohorts proved useful in detecting discontinuities and permitted estimation. The new estimate for the prevalence of ID among the elderly is higher than those previously published, but congruent with the increasing life expectancy of elderly persons with ID.

KEYWORDS

intellectual disability; prevalence; elderly; multiple register study; epidemiology

Introduction

Due to improving health and social welfare, the life expectancy of persons with intellectual disability (ID) is increasing along with that of the general population (Bit- tles et al., 2002; Coppus, 2013; Patja, Iivanainen, Vesala, Oksanen, & Ruoppila, 2000; Perkins & Moran, 2010). Persons with ID have higher mortality than the general population, especially persons with moderate to pro- found ID (Tyrer, Smith, & McGrother, 2007). However, the mortality rate in older age groups, where persons with certain syndromes (e.g., Down) or life-shortening comorbidities have already died, approaches that of the general population (Janicki, Dalton, Henderson, & Davidson, 1999).

Although population-based or cohort studies on the prevalence of ID among the elderly are unavailable, reg- ister-based studies suggest fairly low prevalence figures of less than 0.3% among 70+-year-olds (Kelly, Kelly, & Craig, 2009; Lin, 2009; van Schroyen Lantman-de Valk et al., 2006). This low figure may be due to incom- plete registration of milder forms of ID (Kelly et al., 2009) or other registration problems (Lin, 2009).

Combining multiple registers may provide more com- plete coverage than single registers (Kelly et al., 2009;

Lin, 2009). Our multiple register study of ID found a prevalence of 0.38% among 65+-year-olds. At the age of 65 or 66, however, a sudden drop in the prevalence of ID was recorded from 0.9% to 0.4% (Westerinen, Kaski, Virta, Almqvist, & Iivanainen, 2014). At the age of 65, persons with ID who had previously received a dis- ability pension with a medical diagnosis are reassigned to the old age pension register with no medical diagnosis. For this reason, they can no longer be traced through the main register and only partly via other registers. They are thus effectively lost as ID cases.

Our present study estimates the number of missing 65- to 100-year-olds based on the age distribution of the entire population.

Methods

A list of ID diagnoses served as inclusion criteria, includ- ing only those aetiological diagnoses that typically involve ID (Westerinen et al., 2014). We combined data from the eight national registers described in Table 1. The benefits of the “Disability Pension (DP),” “Disability Allowance (DA),” and “Funding for Rehabili- tation (REH)” registers cease at the age of 65, whereas the

Table 1. The registers used in the study.

Register	Ages	Description
Social Insurance Institution of Finland (KELA)		
Child Disability Allowance	0–15 years	Supports care of disability.
Disability Pension (DP)	16–64 years	Compensates for working incapacity.
Disability Allowance (DA)	16–64 years	Covers costs incurred by mild disability of working-aged persons.
Funding for Rehabilitation (REH)	0–64 years	Mainly for persons with severe disability.
Pensioners' Care Allowance (PCA)	16–64 years with disability pension, 65+ years with retirement pension	Supports home care; provides compensation for extra costs related to disability.
Preferential Refunding of Long-Term Medication (MED)	Any age	Patients with certain long-term diseases are entitled to a 100% refund of medication expenses. One of the indications is behavioural disorders associated with ID.
National Institute for Health and Welfare		
Discharge Register of Hospitals (HOSP)	Any age	1–6 ICD-10 diagnoses are coded at the end of each hospital stay. All persons with an ID diagnosis who had been admitted to hospital during 1996–2000 and were still alive in 2000 were identified.
Discharge Register of Social Care (CARE)	Any age	Persons living in an institution or in housing units, in this case restricted to those with ID.

Note. ID = intellectual disability; ICD-10 = *International Classification of Diseases and Related Health Problems: Tenth Revision*.

“Pensioners’ Care Allowance (PCA),” “Preferential Refunding of Long-Term Medication (MED),” “Discharge Register of Hospitals (HOSP),” and “Discharge Register of Social Care (CARE)” also cover persons 65+ years of age.

Doctors have verified in normal clinical practice the ICD-10 (World Health Organization, 1992) diagnoses of all persons in each register. The application of a benefit is based on a physician’s certificate, and the diagnostic criteria must be fulfilled. These criteria are checked by the Social Insurance Institution of Finland (KELA).

All data were collected in 2000. Because KELA benefits are long term, and to limit the costs of register searching, we limited the sampling in the KELA registers to 1 year.

Finland has a national health insurance scheme funded from tax revenue that covers all 5.18 million residents (in 2000) and is governed by KELA. Each beneficiary has a unique individual social security code, and this identifier served as a registry linkage to prevent double counts. The registers were crosschecked with the Finnish death register to exclude possible deceased individuals. The Finnish national register system is very reliable and includes all and only those persons concerned (Sund, 2012). The risk group consisted of the entire Finnish population of 65- to 100-year-olds ($N = 778,473$) at the end of 2000 (Official Statistics of Finland, n.d.-b).

The sampling from the registers was performed according to the study protocol of the organisations that administer the registers. The investigators had no direct access to the registers and were blind to case identities.

Because of the high reliability of Finnish registers, the prevalence of ID at the age of 65 can serve as a basis for estimating the minimum prevalence of ID. Thus, we corrected the age-specific prevalence of ID based on the total population age distribution so as to eliminate the abrupt drop at retirement age. Age-specific prevalence was adjusted to decrease steadily from the observed average level in the five preceding age cohorts to the level observed in the oldest age groups. We assume that both original observed levels at different ends of the critical age span represent the true minimum level.

The MED register provided an opportunity to test the validity of the results. The MED benefit is assigned according to the same criteria for all ages, and those who receive it can be traced irrespective of age.

We calculated 95% confidence intervals (CI) for the prevalence rates for each cohort group (Gardner & Altman, 1989).

The Ethics Committee of the National Institute for Health and Welfare, Finland, approved the study protocol.

Table 2. The observed and estimated figures for the population with ID over 65 by sex.

		Population with ID 65–100 years			
		Observed		Estimated	
		Number	Prevalence % 95% CI	Number	Prevalence % 95% CI
Total population 65–100 years					
Men	300,174	1,161	0.39% [0.36, 0.41]	2,283	0.76% [0.73, 0.79]
Women	478,299	1,832	0.38% [0.37, 0.40]	3,521	0.74% [0.71, 0.76]
Total	778,473	2,993	0.38% [0.37, 0.40]	5,804	0.75% [0.73, 0.76]

Note. CI = confidence interval.

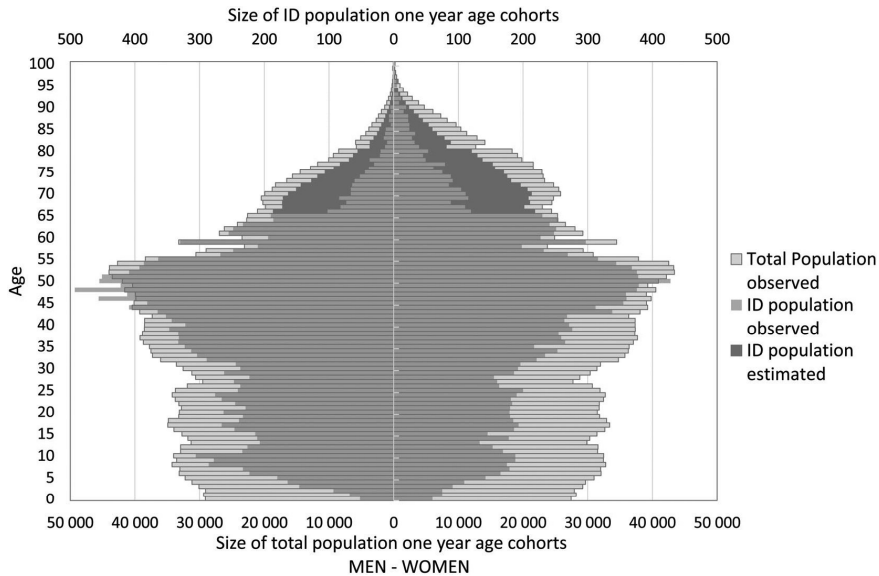


Figure 1. Age distribution of the general Finnish population (bottom scale) and age distribution of the population with ID by sex (top scale). The scale for the population with ID size is 1% of the total population size, so that bars of the same length indicate an ID prevalence of 1% for that age cohort. The observed ID distribution refers to the multiple register study, and the estimated distribution refers to the results in this study. The ID prevalence in the correction was permitted to decrease steadily from the average value observed at ages 61–65, 0.90/0.91% (men/women), to the average value observed in the oldest age groups, 0.30% (both sexes).

Results

Figure 1 shows the age distribution of the entire Finnish population and the observed ID population in the

multiple register study in 2000. The age-specific prevalence dropped from 0.84%/0.91% at age 65 to 0.49%/0.49% at age 66 (men/women). The average prevalence for the five preceding age cohorts prior to the drop was 0.90% for men and 0.91% for women. The average prevalence observed in the oldest age groups (80+ years) was 0.30%. The new estimated figures appear in Table 2.

The correction yielded 2,811 (1,122 men, 1,689 women) new persons with ID, in addition to the 2,993

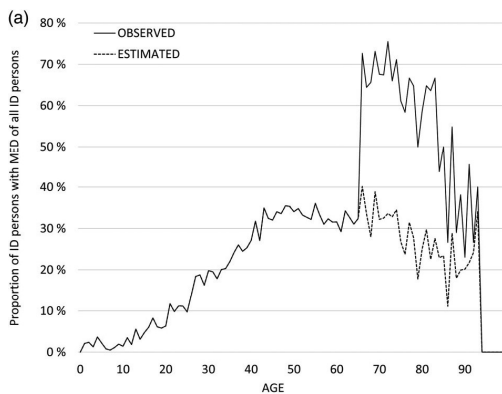


Figure 2a. The proportion of persons who receive the benefit for preferential reimbursement of psychiatric medication for behavioural disorders associated with ID (MED) among all persons with ID by age (Quotient 1). The observed distribution refers to the multiple register study, and the estimated distribution refers to the results in this study. ID prevalence was permitted to decrease steadily from the average value observed at ages 61–65, 0.90/0.91% (men/women), to the average value observed in the oldest age groups, 0.30% (both sexes).

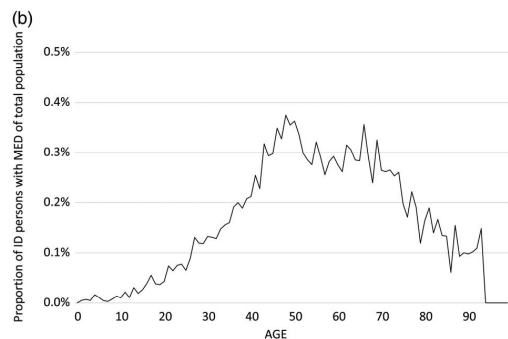


Figure 2b. The prevalence of the benefit for preferential reimbursement of psychiatric medication for behavioural disorders associated with ID (MED) in the total population (Quotient 2).

persons observed in the multiple register study, nearly doubling the number. For 65- to 100-year-olds, the correction yielded a final prevalence of 0.75% (95% CI [0.73, 0.76]), compared to the observed 0.38% (95% CI [0.37, 0.40]).

Of all persons with ID (= Quotient 1), the percentage of individuals receiving the MED benefit rises abruptly from 30% to 70% at retirement age (Figure 2a). In contrast, the percentage of the total population (= Quotient 2) shows no such abrupt change, and the distribution is continuous (Figure 2b). Thus, the abrupt change in Quotient 1 cannot be due to the numerator; rather, it must be due to a diminishing denominator, the total number of persons with ID in each 1-year cohort.

The MED benefit allowance was not connected to retirement age: of 66- to 70-year-olds in 2000, 87% had been granted the benefit over 10 years earlier. This new estimate of elderly persons with ID, which served as the denominator in Quotient 1, rendered the distribution continuous (Figure 2a).

Even a rise in the mortality rate among 75+-year-olds would not substantially influence the new total prevalence estimate of all old ages combined, since 63% of "new cases" were 66- to 75-year-olds. Applying the correction only to this age span would raise the total prevalence among 65- to 100-year-olds to 0.61%.

Discussion

The new estimated ID prevalence based on population structure doubled the number of persons with ID over 65 years and resulted in a prevalence rate of 0.75%. The noted loss of older cases in the multiple register study, which was due to a loss of diagnostic information in the registers at the retirement pension age (65 years), seems to have large practical significance.

The observed drop in the age-specific prevalence rate among 65+-year-olds was corrected according to the Finnish population structure in order for it to decrease steadily from the average level observed in the preceding five age cohorts to the average level observed in the oldest age groups. However, the end level may also be an underestimation due to insufficient register coverage of 65+-year-olds. Raising the correction level at the far end would influence figures immediately after the age of 65 years only slightly. Although mortality decreases the prevalence along with age, it cannot happen suddenly at this age point. As was noted earlier, even a limited correction only for the age cohorts of 66- to 75-year-olds immediately beyond the discontinuity point would raise the new estimate to 0.61%. This estimate is much higher than those reported in previous studies of 65+-year-olds

with ID (Kelly et al., 2009; Lin, 2009; van Schroyen Lantman-de Valk et al., 2006).

The validation method in our multiple register study was based on the MED register. The abrupt increase in the proportion of MED benefit receivers observed in all persons with ID is not due to the different allowance criteria for this benefit for individuals around the age of 65, as prevalence of the benefit calculated for the total population was continuous. Furthermore, this period has seen no substantial changes in the birth rate (Official Statistics of Finland, n.d.-a) or in the mortality of persons with ID (Patja et al., 2000). The cause of the observed discontinuity may thus reliably stem from the dropping of older persons with ID from the registers.

Some researchers have expressed concern about the lack of developed services for persons with ID (Holt et al., 2000; Whitaker, 2004). The modern trend to include persons with ID in mainstream services may lead to an unwillingness to diagnose a personal ID or to register it for fear of stigmatisation (Fryers, 1987). At the local service provision level, persons with ID should be recognised, because they are seldom able to express their personal needs themselves. At the governmental level, reliable knowledge of ID prevalence should form the basis for allocating sufficient resources for organising appropriate services.

To our knowledge, few previous studies have explored the prevalence of ID among the elderly (Maulik, Mascarenhas, Mathers, Dua, & Saxena, 2011). Our study was based on eight national registers. However, the key register (DP) for working-age persons becomes unavailable at retirement age (65 years), resulting in a sharp drop in the prevalence of ID. Finland has a long tradition of comprehensive censuses, which made it possible to calculate estimates beyond the discontinuity point. The possibility of using one of the registers to validate the new estimates strengthened the validity of our results. It is also worth noting that all citizens in Finland, regardless of their socioeconomic status or place of residence, are entitled to and enjoy equal access to health services (Niemelä & Salminen, 2006).

Our study is subject to the same limitations that apply to register-based studies of the prevalence of ID. Inclusion in a sample depends on numerous administrative decisions. This type of study informs us as much about how a society functions as it does about the prevalence of a condition, even when the condition is well defined (Fryers, 1987). This discontinuity in the prevalence of ID at age 65 is specific to Finland, so the estimation approach used here may be inapplicable to other countries. However, comparable problems in register coverage may exist elsewhere. For example, milder forms of ID may be missing, thus lowering the

prevalence rates of ID among the elderly (Kelly et al., 2009). The estimation procedure was based on certain assumptions; using other plausible estimation procedures would have resulted in different estimates.

Conclusions

A high number of elderly persons with ID are missing due to registration criteria in the multiple register study. To correct for this deficit, we applied a method based on population age structure. This improved method yielded prevalence estimates that were higher than those reported in previously published studies of elderly persons with ID. This finding should undergo testing in other contexts.

Conflicts of interest

The funding bodies have imposed no restrictions on free access to or publication of the research data. The authors have no conflicts of interest, whether financial or nonfinancial, and receive no financial benefit, whether direct or indirect.

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The nationwide register-based prevalence of intellectual disability during childhood and adolescence

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Abstract

Background Many studies have evaluated the prevalence of intellectual disability (ID) by focusing on different ages during childhood and adolescence. Although the prevalence of ID is higher in older age groups, how cumulative prevalence increases, and what level it reaches before adulthood, remains unclear.

Method We used Care Register for Health Care to retrieve information on individuals born in 1996–2007 with any of the inclusion diagnoses of ID (F7 group and/or aetiological diagnoses) for the period 1996 to 2013. The cumulative prevalence was calculated as percentages for every age based on Finnish population data.

Results The registration of new diagnoses of ID continued steadily throughout the developmental years. The cumulative prevalence reached 1.19% by age 17.5 among those born in 1996. Later-born age groups appeared to receive their first ID diagnoses earlier in childhood. Those born in 1999 reached a

cumulative prevalence of 1.21% already by age 14.5. Of all those with ID, 67% had an F7 diagnosis only, 42% had an aetiological diagnosis only and 9% had both diagnoses.

Conclusions Cumulative prevalence of ID by year, until the age of 18, will provide a better estimate and understanding of the prevalence of ID than a point prevalence at any one point during the developmental years.

Keywords adolescent, child, epidemiology, intellectual disability, prevalence, registries

Introduction

Intellectual disability (ID) is, by definition, a condition involving a reduced level of intellectual functioning that results in an individual's diminished ability to adapt to the daily demands of a typical social environment (WHO 1996). A further requirement is that the condition manifest during the individual's developmental period, before the age of 18.

The condition may appear at any age, from early prenatal life throughout childhood and adolescence until adulthood, as a result of acute or progressive

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diseases affecting brain function (genetic syndromes, intrauterine infections, birth complications, subsequent infections, head traumas, drowning accidents, poisonings, etc.) or an individual's apparent inability to comply with increasing normative demands, which then warrants diagnostic investigations. While the condition of ID virtually never disappears once reliably diagnosed, an initial diagnosis at any age will mark the beginning of a cumulative prevalence, which peaks by the end of the developmental period. Mortality among those with severe ID is markedly higher than in the normal population, especially in early childhood, but mortality of children with mild ID may not differ from that of normal children (Similä *et al.* 1987; Patja *et al.* 2000; Hatton *et al.* 2014).

Only two reviews have described studies on the prevalence of ID during childhood and adolescence (Roeleveld *et al.* 1997; Maulik *et al.* 2011). The studies used varying definitions of ID and placed different emphases on the limitations of adaptive behaviour besides intellectual functioning; some of the studies used an IQ cut-off value of 75 for mild ID instead of 70, the ICD-10 value. Of the studies, 80% were cross-sectional, reporting on populations with wide and varied age ranges (Maulik *et al.* 2011). The cohort studies provided higher estimates of ID prevalence than did the cross-sectional studies. The studies that used community-based household sampling and psychometric scales provided higher prevalences than did those that used hospital data/administrative registers and/or official ICD or DSM diagnostics. The studies that took place in developing countries or among low-income populations had higher prevalences than did those in higher-income countries or among more affluent populations (Maulik *et al.* 2011).

The prevalence of severe ID (sID), where the intelligence quotient (IQ) is less than 50, has remained rather stable across studies, peaking at 0.3–0.4% by the age of 18 (Roeleveld *et al.* 1997). On the other hand, the prevalence estimates of mild ID (mID, IQ 50–70) have varied widely, from less than 0.5% up to 10% (Roeleveld *et al.* 1997; Simonoff *et al.* 2006; Emerson 2012). Most cohort studies in the meta-analysis (Maulik *et al.* 2011) showed prevalences of ID in childhood and adolescence between 0.6% and 1.4% (Blomquist *et al.* 1981; Baird & Sadovnick 1985; Hagberg *et al.* 1987; Kääriäinen 1987; Murphy

et al. 1995; Patja *et al.* 2000; Heikura *et al.* 2003; Petterson *et al.* 2007). Two cohort studies showed a much higher prevalence, both 3.7%. One was based on voluntary joining to a long-term Collaborative Perinatal Project (Camp *et al.* 1998), the other was from Lahore, Pakistan from urban slum/mixed urban–rural population (Gustavson 2005).

Most prevalence studies focusing on childhood/adolescence have been cross-sectional, reporting on populations with wide and varied age ranges, and presenting the average prevalence in those age ranges (Maulik *et al.* 2011). However, we could find no study that provided the prevalence of ID by yearly categories throughout developmental ages.

The aim of this study was to estimate the cumulative prevalence of ID throughout childhood and adolescence in Finland based on data in the nationwide register.

Methods

In Finland, diagnostic assessments of ID are mostly conducted in hospitals, outpatient clinics or inpatient wards by specialists in paediatrics, child neurology or child psychiatry.

In this study, we used the Care Register for Health Care (CRHC) in Finland. The CRHC is a continuation of the Hospital Discharge Register, which contains data on patients discharged from hospitals between 1969 and 1993. Since 1994, the CRHC has also included visits in specialised medical outpatient care (National Institute for Health and Welfare n.d.). A review of 32 studies comparing the quality of the CRHC register to external information evaluated its accuracy as good (Sund 2012). Other studies have evaluated the validity of the diagnoses in the CRHC as good, especially for two childhood neurodevelopmental disorders: childhood autism and Tourette's syndrome (Lampi *et al.* 2010; Leivonen *et al.* 2014).

We searched the CRHC for all patients born in 1996–2007 with the inclusion diagnoses of ID. The list of ICD-10 diagnoses included the group F70–F79 (diagnoses of the level of ID) and aetiological diagnoses that typically indicate ID (see Appendix).

The CRHC records treatment periods in specialised medical care as well as the diagnoses at the end of each period (at patient discharge). Those born

in 1996 could be followed-up until the age of 17.5, whereas those born later, only for a shorter period.

Finland has a comprehensive registration system for all its inhabitants that is governed by the Population Register Centre in Finland (Population Register Centre n.d.). Each person has a unique individual personal identity code, which we used to prevent double counts. We cross-checked the CRHC with the Finnish death register to identify individuals who may have died before 1 January 2014. We used data from Statistics Finland (Official Statistics of Finland n.d.) to calculate the prevalence of ID in the Finnish population for each year and age group.

We defined the first occasion of any ID diagnosis for each person. We then calculated the number of people with the first ID diagnosis for each birth year and age. Then, we calculated the cumulative number for each age by adding together the first occurrences until that age, and separately for each group with the same birth year. To calculate the cumulative prevalence, we divided the cumulative number by the number of children alive at the same age at the end of that year. Thus, we excluded those who had died in earlier years from the calculation for that year, from both the index cases and the general population. The prevalence figure for our study does not represent a birth cohort figure, because we did not define our study population based on its year of birth. Instead, the prevalence figure represents the cumulative prevalence of people in the population of Finland with ID at different ages throughout the study years.

We calculated the prevalence figures as average percentages of three succeeding years to smoothen the annual random variation. We calculated 95% confidence intervals (CI) according to Gardner and Altman (1989).

Results

The sampling identified 7975 people diagnosed with any of the ID inclusion diagnoses at least once. Of these, 4826 (61%) were boys. Of those diagnosed with ID, 360 (4.6%) died during the period 1996–2013. Of those born in 1996–1998 and followed up until the ages of 15.5–17.5 (2239 people with ID) 91 (4.1%) had died.

New diagnoses of ID were registered steadily throughout the developmental years, somewhat more often between the ages of 4 and 7 (see Table 1). Those born in 1996 could be followed up until the age of

17.5, when the prevalence reached 1.19% (95% CIs [1.11, 1.28]): 1.42% (95% CIs [1.29, 1.55]) for boys and 0.95% (95% CIs [0.84, 1.06]) for girls. Those born in 1999 already reached this prevalence (1.21%) by the age of 14.5 (95% CIs [1.12, 1.30]) (Fig. 1). When we compared those born in 1996–1998, 1999–2001, 2002–2004 and 2005–2007, the cumulative prevalence by 4.5 years-of-age was roughly the same (0.46–0.49%, Fig. 2). Beyond that age, however, the registration of ID diagnoses became more frequent in later-born age groups. By the age of 6.5 years, the prevalence of ID was 0.66% (95% CIs [0.62, 0.70]) for those born in 1996–1998, but was 0.80% (95% CIs [0.76, 0.84]) for those born in 2005–2007.

The effect of cumulative searching throughout the developmental years was influential compared with point prevalences. When the search was performed for one year only, the figure was on average 0.30% (maximum 0.58%) depending on the age and year of searching.

The impact of mortality on the final cumulative prevalence was rather small. For those born in 1996 and followed up until the age of 17.5, without deaths the final cumulative prevalence would have been 1.23% instead of 1.19%.

By the end of the follow-up period, 5506 (72.3% of all) people had an F7 diagnosis, 2698 (35.4%) had an aetiological inclusion diagnosis and 670 (8.8%) had both an F7 and aetiological diagnosis. Among those with only an aetiological diagnosis and no F7 diagnosis (altogether 2028 people), the most frequent diagnosis was Down's syndrome; only 10.1% of 929 people with Down's syndrome held an F7 diagnosis.

We calculated the distribution of diagnoses at the developmental level for those born in 1996–1998 and followed up to the age of 15.5–17.5 years. In this group, 1609 (75%) of 2148 ID people had an F7 diagnosis, providing a population prevalence of 0.94% in 2013 (see Table 2). Of those with an F7 diagnosis, 1266 (79%) had a defined level of ID. The population prevalence of mID was 0.52% and of sID, 0.21%; for 0.20%, the developmental level was undefined (ICD-10 diagnosis F78.0–9 or F79.0–9).

Discussion

In our study, the cumulative prevalence of ID reached 1.19% by age 17.5 among those born in 1996.

Interestingly, the prevalence increased rather steadily

Table 1 New diagnoses and cumulative prevalence of ID by year of birth and average age

New registrations of ID diagnoses by year of birth (vertical) and average age (horizontal)																		
	0.5	1.5	2.5	3.5	4.5	5.5	6.5	7.5	8.5	9.5	10.5	11.5	12.5	13.5	14.5	15.5	16.5	17.5
1996	84	41	53	45	40	60	63	43	35	27	20	23	42	41	38	43	42	33
1997	89	70	52	53	48	57	67	36	25	21	24	22	21	39	34	44	53	
1998	99	51	41	43	50	54	62	58	30	26	43	37	29	32	33	23		
1999	111	54	45	41	44	72	87	45	40	23	44	29	48	39	32			
2000	117	37	49	30	58	68	73	52	36	44	44	40	44	25				
2001	112	40	37	38	58	63	79	59	49	36	46	36	35					
2002	117	44	35	43	47	75	70	54	53	50	40	27						
2003	95	41	33	37	73	82	76	81	33	31	31							
2004	97	42	36	49	53	98	93	78	55	33								
2005	118	40	50	53	52	91	107	51	45									
2006	123	36	49	51	61	89	77	64										
2007	115	54	33	50	58	103	105											
2008	122	48	35	49	70	100												
2009	122	50	31	34	69													
2010	81	51	40	64														
2011	105	55	46															
2012	102	46																
2013	100																	
Cumulative prevalence by year of birth (vertical) and average age (horizontal)																		
	0.5	1.5	2.5	3.5	4.5	5.5	6.5	7.5	8.5	9.5	10.5	11.5	12.5	13.5	14.5	15.5	16.5	17.5
1996	0.14%	0.19%	0.28%	0.35%	0.41%	0.51%	0.61%	0.67%	0.73%	0.77%	0.80%	0.83%	0.89%	0.96%	1.01%	1.08%	1.14%	1.19%
1997	0.14%	0.26%	0.34%	0.42%	0.50%	0.59%	0.71%	0.76%	0.80%	0.83%	0.87%	0.91%	0.93%	0.99%	1.05%	1.12%	1.20%	
1998	0.17%	0.25%	0.32%	0.39%	0.47%	0.57%	0.67%	0.76%	0.81%	0.85%	0.92%	0.98%	1.03%	1.08%	1.13%	1.16%		
1999	0.18%	0.27%	0.35%	0.41%	0.49%	0.60%	0.75%	0.82%	0.88%	0.92%	0.98%	1.03%	1.11%	1.16%	1.21%			
2000	0.18%	0.24%	0.32%	0.37%	0.47%	0.58%	0.70%	0.79%	0.85%	0.92%	0.99%	1.06%	1.13%	1.17%				
2001	0.19%	0.26%	0.32%	0.38%	0.49%	0.60%	0.73%	0.83%	0.91%	0.97%	1.05%	1.10%	1.16%					
2002	0.19%	0.27%	0.33%	0.40%	0.48%	0.61%	0.73%	0.82%	0.90%	0.98%	1.05%	1.09%						
2003	0.14%	0.20%	0.26%	0.32%	0.45%	0.59%	0.71%	0.85%	0.90%	0.95%	1.00%							
2004	0.15%	0.22%	0.28%	0.35%	0.44%	0.60%	0.75%	0.87%	0.96%	1.01%								
2005	0.18%	0.24%	0.32%	0.41%	0.49%	0.64%	0.82%	0.90%	0.96%									
2006	0.19%	0.24%	0.32%	0.40%	0.50%	0.64%	0.76%	0.86%										
2007	0.18%	0.27%	0.32%	0.40%	0.49%	0.66%	0.83%											
2008	0.19%	0.27%	0.33%	0.41%	0.52%	0.67%												
2009	0.19%	0.27%	0.31%	0.36%	0.47%													
2010	0.13%	0.20%	0.26%	0.36%														
2011	0.16%	0.25%	0.32%															
2012	0.15%	0.22%																
2013	0.17%																	
1996-1998	0.15%	0.23%	0.31%	0.39%	0.46%	0.56%	0.66%	0.73%	0.78%	0.82%	0.86%	0.91%	0.95%	1.01%	1.06%	1.12%	1.17%	1.19%
1999-2001	0.18%	0.25%	0.33%	0.39%	0.48%	0.59%	0.73%	0.81%	0.88%	0.94%	1.01%	1.06%	1.13%	1.17%	1.21%			
2002-2004	0.16%	0.23%	0.29%	0.36%	0.46%	0.60%	0.73%	0.85%	0.92%	0.98%	1.02%	1.09%						
2005-2007	0.18%	0.25%	0.32%	0.40%	0.49%	0.65%	0.80%	0.88%	0.96%									

throughout the developmental age, with the most rapid increase occurring between ages 4 and 7. The prevalence reached 1.21% already by age 14.5 among those born 1999. Only three in four people with ID had an F7 diagnosis; we identified the others only by their aetiological diagnoses.

To the best of our knowledge, this is the first study to provide cumulative prevalence figures for various ages throughout childhood. These figures demonstrate how diagnostics in 'real-life' health care systems add up to the final prevalence of ID.

This study carries several strengths. First, every Finnish citizen has a unique personal identification code, which remains the same throughout his or her lifetime, which makes a register study easy and reliable. Second, we could use one national register of specialised care, the CRCH, instead of several other registers or other tedious methods for case finding. In Finland, social and health benefits for ID people require an ICD diagnosis, which is made during investigations in specialised care. Thus, CRCH registration represents a gateway for ID patients to

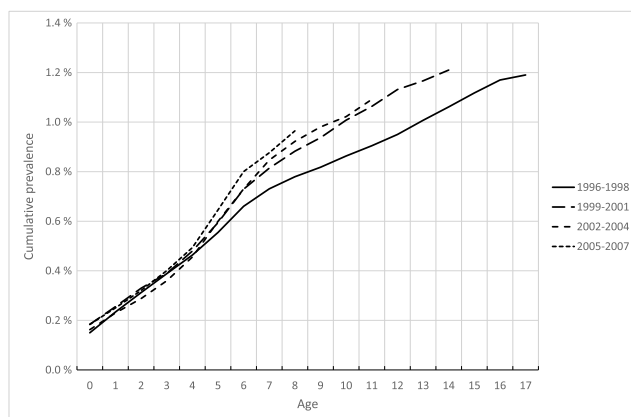


Figure 1 Cumulative prevalence of ID at different ages for four consecutive age classes born 1996–1998, 1999–2001, 2002–2004 and 2005–2007.

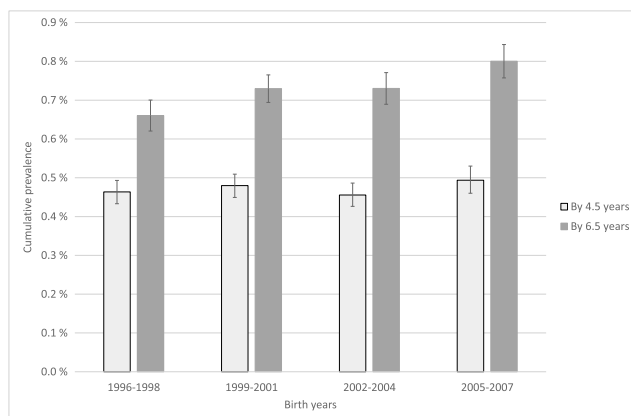


Figure 2 Cumulative prevalence of ID by ages 4.5 and 6.5 years in four consecutive age classes with 95% confidence intervals.

receive further assistance and benefits. Third, cumulative searching mirrors the actual role of this register: people may visit specialists for diagnostic and check-up purposes only at some ages, after which the original diagnosis prevails. Finally, studies have found the accuracy of the CRHC to be reliable when compared to corresponding individual records (Sund 2012).

The most important limitation of our study is the inclusion in our ID prevalence of only diagnoses which regularly indicate ID. Because many other diagnoses, such as tuberous sclerosis, hydrocephaly and serious infections of the central nervous system, among others, can sometimes be associated with ID, our figures probably underestimate the true

prevalences. Another limitation of our study is the lack of register data on the level of ID. Only 60% of those with ID had a defined level.

Our main result, a cumulative prevalence of 1.19% at the end of the developmental period, is somewhat higher than the prevalence in the cumulative meta-analysis (Maulik *et al.* 2011), where the estimates stabilised at around 1.1% in the studies published between 2000 and 2008. Compared to cross-sectional studies and to those based on DSM/ICD diagnostics, however, the difference is even greater.

In those born in 2002, the cumulative prevalence by age 11.5 rose to the same level (1.09%) as the prevalence figure in two Northern Finland birth

Table 2 Distribution of developmental levels in people with ID born in 1996–1998, registered by the end of 2013. F7 refers to the ICD-10 diagnostic category of intellectual disability. mID, mild ID; sID, severe ID in the meaning of combining levels that are more severe than mild

TOTAL	F7 diagnosis	Defined level	Mild	mID
2148	1609 (75%)	1266 (79%)	909 (72%)	
		Undefined level	Moderate	sID
		343 (21%)	231 (18%)	(28%)
	No F7 diagnosis		Severe	
	539 (25%)		83 (7%)	
			Profound	
			43 (3%)	

cohort studies (those born in 1966 and in 1985–1986) by age 11.5 (Heikura *et al.* 2003). This may indicate that the Finnish health care system generally finds the cases as thoroughly as a well-planned prospective cohort study. The birth year in the second cohort study differs from that in the cohort in our study by more than ten years, which may have led to changes in the incidence of ID. On the other hand, the prevalence of ID in Northern Finland during the 30 years between the two cohort studies remained stable.

A previous study also used the CRHC to define the prevalence of ID at the age of 14 for those born in 1996 (Gyllenberg *et al.* 2014). That study searched only for F7 diagnoses and not aetiological diagnoses. Their lower figure (0.7%) is thus in line with ours.

The rapid increase in cumulative prevalence observed between ages four and seven indicates an active screening for developmental problems at that age in the Finnish child health care system (Valtonen *et al.* 2004). The registration of ID diagnoses became more frequent among later-born samples, most probably because of a gradual increase in the nationwide use of screening instruments for developmental problems at the age of four. In 2007, screening covered 84% of all child health care centres (Hakulinen-Viitanen *et al.* 2008). Another possibility is a real increase in the incidence of ID, but the evaluation of this hypothesis is beyond the scope of the present study.

Of those with ID, 75% had an F7 diagnosis; others were identified only by their aetiological diagnoses. Of those with a defined level of ID, 72% had mID. If the

distribution by ID level were the same for those without a defined developmental level (also including those with only an aetiological diagnosis), the population prevalence would be 0.85% for mID and 0.34% for sID. The latter figure is in line with those of previous studies (Roeleveld *et al.* 1997).

F7 diagnoses were recorded in only 22% of cases with an aetiological diagnosis. The proportion was even less in those with Down’s syndrome (8.5%), which clearly reveals some systematic tendency. The reason may be that some consider an F7 diagnosis unnecessary for Down’s syndrome or other syndromes with ID. This may lead to the postponement and assignment of the responsibility of defining the person’s developmental level to the educational system or to programs that provide special care for ID people.

The general health care register CRHC can be useful for estimating the prevalence of ID. Analysing the first occurrences in the register, and prevalence by age, will provide feedback on the effectiveness of screening for developmental problems. However, Finnish health care systems do not systematically register individuals’ developmental levels.

Our study shows the importance of paying attention to the age at which the prevalence of ID is evaluated. The final cumulative prevalence at the end of the developmental period indicates what proportion of the adolescent population will require special services related to ID when they reach adulthood. The average prevalence of ID across all childhood/adolescence years may be misleading.

Because needs for rehabilitation, support and services at different ages vary, average estimates may be of little use.

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- Appendix A: List of inclusion diagnoses.**
- | | | | |
|-----------------|--|---|--|
| E72.5 | Disorders of glycine metabolism | F71.* | Moderate mental retardation |
| E75.0,1,2,4,6 | Disorders of sphingolipid metabolism and other lipid storage disorders | F72.* | Severe mental retardation |
| E76.0,1,2,3,8,9 | Disorders of glycosaminoglycan metabolism | F73.* | Profound mental retardation |
| E77.0,1,8,9 | Disorders of glycoprotein metabolism | F78.* | Other mental retardation |
| E79.1 | Lesch–Nyhan syndrome | F79.* | Unspecified mental retardation |
| E88.8 | Other specified metabolic disorders | F84.2 | Rett's syndrome |
| F70.* | Mild mental retardation | Q00.0 | Anencephaly and similar malformations |
| | | Q03.81 | Hydrolethrus syndrome |
| | | Q04.00, | Other congenital malformations of the brain |
| | | Q04.2, Q04.51 | Foetal alcohol syndrome (dysmorphic) |
| | | Q86.0 | Chromosomal microdeletions, specified syndromes |
| | | Q87.01,05,12,-15,18,19,23,30-31,38,83–6 | |
| | | Q90.* | Down's syndrome |
| | | Q91.* | Edwards' syndrome and Patau's syndrome |
| | | Q92.*, except Q92.6 | Other trisomies and partial trisomies of the autosomes, not classified elsewhere |
| | | Q93.* | Monosomies and deletions from the autosomes, not classified elsewhere |
| | | Q99.2 | Fragile X syndrome |

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